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ABSTRACT

This report, arising from a 1991 meeting, provides an update to two of the six areas covered in the 1989 long-term plan of the National Institute on Deafness and Other Communication Disorders. These include: (1) balance and the vestibular system; and (2) language and language impairments. For each area, the state of the art is reviewed, recent accomplishments summarized, and program goals and research opportunities listed. Subcommittees in these areas examined the Institute's research portfolio, identified changes in the field, recommended levels and areas of research activity, and suggested potential initiatives. Among 11 areas of recommended research in vestibular function are: signal transduction by vestibular end-organs; sensory integration in spatial orientation, perception and motion sickness; development and aging of the vestibular system; prevalence and environmental factors; diagnostic methods and testing procedures; and adaptive mechanisms. Areas of recommended research in language include: multicultural issues; language and deaf people (language development in deaf children, studies of literacy in deaf children and adults, and sign language structure and function); language disorders in children (bases of language disorders, assessment, academic and social/vocational impact, and intervention); and language disorders in adults (brain-language relations, processes underlying language disorders, assessment and intervention, and comparative language). Appendices include Public Law 100-553 (the National Deafness and Other Communication Disorders Act of 1988) and a listing of panel members. (DB)

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National Institute on Deafness and
Other Communication Disorders

National Strategic Research Plan

for
Balance and the Vestibular System

and
Language and Language Impairments

1991

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FOREWORD

On October 28, 1988 the National Deafness and Other Communication Disorders Act of 1988 became Public Law 100-553 (Appendix A) establishing the National Institute on Deafness and Other Communication Disorders (NIDCD) within the National Institutes of Health (NIH). The law required that the Director, NIDCD, establish a National Deafness and Other Communication Disorders Program and prepare a plan to initiate, expand, intensify and coordinate Institute activities concerning disorders of hearing, balance, smell, taste, voice, speech and language.

In response to this mandate, in January 1989 a Task Force of scientific experts representing the seven program areas of the Institute prepared the first National Strategic Research Plan which has guided the Institute in its research activities over the last two years. The National Strategic Research Plan is also intended to inform the Nation's scientists of areas of opportunity for research and to provide them with guidance as they formulate their own research plans. The plan informs persons with communication disorders and their support organizations of past research accomplishments and potential future activities. In addition, the plan is intended to inform members of Congress of research progress and future research opportunities in scientific areas within the purview of the NIDCD.

P.L. 100-553 requires the National Deafness and Other Communication Disorders Advisory Board to review, evaluate and update the plan periodically to assure its continuing relevance. To meet this legislative mandate, the National Advisory Board in the spring of 1990 decided that it would update two of the six sections of the Plan every year thus updating the entire plan within a three-year period. The two sections the National Advisory Board selected to update in 1991 were Balance and the Vestibular System and Language and Language Impairments. The National Advisory Board established subcommittees for these two areas which met and made recommendations for panel members to update the Balance and the Vestibular System and Language and Language Impairment sections of the plan, compared the research portfolio of the Institute to the National Strategic Research Plan, identified changes in the field since the plan was developed, recommended levels and areas of research activity and suggested potential initiatives. On January 28-29, 1991 panels of scientific experts in the areas of balance and the vestibular system and language and language impairment were convened. (Members of the panels are listed in Appendix B.) The results of these meetings are contained in this report.

Members of the panels are due special thanks for giving generously of their talents and time in seeing this document through to completion. Their meetings on

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January 28 and 29, 1991 brought together representatives of a broad array of scientific disciplines within the areas of balance and the vestibular system and language and language impairments. Members of the panels shared their diverse ideas and worked diligently to achieve consensus on a comprehensive view of each field and a vision for the future. Subsequently, panel members refined their efforts with numerous revised texts.

Dr. Horst R. Konrad and Dr. Barry W. Peterson, Cochairpersons of the Balance and the Vestibular System Panel, and Dr. Rita S. Berndt and Dr. Laurence B. Leonard, Cochairpersons of the Language and Language Impairment Panel, should be singled out for special appreciation. This update to the National Strategic Research Plan is very much a product of their experience, expertise and guidance. Joined by their fellow panel members, they have formulated a plan for future research in two important scientific areas of the NIDCD.

James B. Snow, Jr., M.D.
Director
National Institute on Deafness and
Other Communication Disorders

PREFACE

The National Strategic Research Plan of the National Institute on Deafness and Other Communication Disorders (NIDCD) was prepared in April 1989 and presented the research recommendations of more than 100 eminent scientists in the seven program areas of the NIDCD. These areas are hearing, balance, smell, taste, voice, speech and language. This report contains an update of the balance and the vestibular system and language and language impairments sections of the 1989 plan.

Although disorders of balance and language affect different populations, both have a dramatic impact on the ability of people to live full and productive lives. The vestibular system maintains balance and posture, regulates locomotion and other volitional movements and provides a conscious awareness of orientation in space and a visual fixation in motion. Disease, aging, exposure to unusual motion or altered gravitational environments can impair balance. A major consequence of vestibular disturbance is diminished capability and desire for purposeful activity. A better understanding of the normal function, development and aging of the vestibular system and the disorders which affect it would improve national health and reduce future health care costs.

Language is the uniquely human means of communication through which knowledge, belief and behavior can be explained and shared. The ability to manipulate language to satisfy needs and desires and to express thoughts, observations and values is an important human pursuit that directly influences the quality of life for any individual. Language impairments impede economic self-sufficiency, academic performance and employment opportunities. In addition to loss of livelihood, these disorders impose social isolation and personal suffering on the affected individuals and place an enormous emotional and economic burden on their families and on society as a whole.

Language and balance disorders have long been recognized but their causes have not been well understood. Current advances in scientific knowledge and technology now make it possible to make unprecedented progress in these long neglected problems.

PREFACE

This update of the balance and vestibular system and language and language impairment sections of the National Strategic Research Plan has provided an opportunity to evaluate the progress that has been made in these areas during the first few years of the Institute and to assess future needs and opportunities. The recommendations presented here are expected to lead to exciting advances in our scientific knowledge and improve the quality of life for many people.

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Cochairpersons, Balance and
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**National Institute on Deafness and
Other Communication Disorders**

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Balance and the Vestibular System

Over 90 million Americans have experienced dizziness or a balance problem. The cost of medical care for patients with balance disorders exceeds one billion dollars per year. Balance disorders increase in frequency in the older age groups and by age 75 years become the most common reason for seeking help from physicians. Over 50% of elderly patients interviewed at home complain of balance disorders. When one adds the cost of injury caused by imbalance and the lives and aircraft lost by the armed services due to balance-related disorientation, the cost becomes truly staggering. Since our population is aging, more Americans will suffer from balance disorders in the future. A better understanding of the normal function, development and aging of this system and the disorders which affect it would certainly improve national health and reduce future health care costs. The need to understand motion sickness and orientation in high technology aircraft and altered gravity environments is also pressing. Our nation increasingly relies on such aircraft for national defense and

has reaffirmed major space exploration as a national goal.

The primary receptors of the vestibular system are located in the inner ear and the system is required for the sense of orientation in space, for maintaining balance and posture, for regulating locomotion and fine motor control and for the ability of a person to fix in view and follow an object when the object or the person is in motion. The brain also integrates information from the vestibular detectors and their central connections with information from the eyes, skin, muscles and joints to create perception of body orientation and of motion of the body with respect to space. Because the system has many components, there are many symptoms that reflect its malfunction.

A current view of the vestibular system is that the vestibular receptor organs "process" the forces associated with head acceleration and changes in head position relative to gravity. Nerves going to brain control centers are excited, and the brain uses these signals to develop a sense of orientation and to activate automatically the muscles that subserve movement of the eyes,

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locomotion and posture. Many of the vestibular pathways in the central nervous system are organized into reflex pathways that serve to stabilize and coordinate movements of the eyes, head and body. These processes are collectively referred to as the vestibular reflex systems. One such reflex, the vestibulo-ocular reflex, causes the eyes to move in reaction to head motion.

Orientation is a complex function that allows a person to know the relationship of parts of the body to the environment in a dynamic and reciprocal interaction. Maintaining orientation and balance requires the integration of information from many sources, especially the ears, eyes, skin, muscles and joints, and transforming these signals into coordinated patterns of muscle activity. Because vestibular signals interact with all the major sensory systems and involve major brain centers, a large number of disease processes can impair balance.

Motion sickness can occur with exposure to conflicting orientation cues, perceptions of space and motion signals. A high incidence of motion sickness occurs in pilots being trained in stationary flight simulators. Trainees learn behavioral responses to prevent the development of motion sickness. Space motion sickness, which affects a large number of astronauts, is a major unsolved problem for space travel.

During the past decade, new protocols and computer-based techniques have been developed that can help in the evaluation of gaze and balance disorders. Vestibular reflexes, posture and the interactions of different sensory inputs to the central nervous system can be measured precisely. Among the most remarkable properties of the vestibular and balance control systems are their ability to maintain useful functioning responses to many novel motion environments and their ability to adapt to abnormal function in one or more of their components. Recent accomplishments include a recognition of the essential role of adaptive plasticity in the normal function of the vestibular system, development of improved mathematical models of vestibular reflexes and an anatomical description of the basic circuitry in the brain stem subserving the vestibular-ocular reflexes. Important advances have also been made in understanding the structure and function of the vestibular receptor organs and the way in which their activity is modulated by efferent signals originating in the brain. Work is now beginning to determine the neurotransmitters utilized by neural circuits in the vestibular system, to define the changes that occur in this system during development and aging and to devise efficient techniques for rehabilitating patients with vestibular dysfunction.

Advances in diagnosis include computer-based technology to identify, quantify and locate the source of balance

dysfunction. The National Research Council recently suggested methods for standardization of testing, test interpretation and technician training. Ongoing improvement in magnetic resonance imaging provides for detection of smaller, more operable neoplasms and the detection of several degenerative brain disorders including cerebral atherosclerosis, multiple sclerosis and spinocerebellar disorders. These developments have been complemented by the ability of dynamic computed tomography to measure blood flow in various parts of the brain. Such techniques result in early diagnosis and improve the outcome of surgery. They also allow us to evaluate better the progression of disease, design clinical research programs and evaluate the effects of treatment. Some autoimmune disorders and ataxic syndromes can now be diagnosed and treated with medication based on a better understanding of immune mechanisms and the role of brain transmitters. There are recent improvements in our understanding of the association between balance symptoms and the social, psychological and psychiatric disorders which frequently accompany them. This understanding allows us to manage these patients in a comprehensive fashion rather than to focus on isolated balance disorders.

Major Basic Scientific Opportunities

There are five broad, high-priority areas for basic vestibular research: signal transduction by vestibular end-organs; reflex control of posture and gaze; sensory integration in spatial orientation, perception and motion sickness; adaptive changes in vestibular function; and development and aging of the vestibular system. Within each area, opportunities exist for research at the levels of: behavior, kinesiology and biophysics; anatomy and physiology; cellular properties, biophysics and metabolism; pharmacology, molecular biology and genetics; and mathematical and computer modeling. These opportunities can be summarized as follows:

Signal Transduction by Vestibular End-Organs

Important goals are to understand how vestibular end-organs convert head movements into neural signals and to determine the way in which this conversion is modulated by efferent projections from the brain to the end-organs. Research is needed to:

- o Describe the motions of the cupulae and otolithic membranes in response to head movements and examine the possible role of hair cell motility in modifying these motions.
- o Determine the distribution and organization of otolithic organ and semicircular canal inputs to the

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vestibular nuclei and cerebellum and elucidate the function of the vestibular efferent system in behaving animals through anatomic and physiologic research.

- o Characterize the biophysical properties of hair cells and afferent fiber terminals. This research should include studies of the hair cell transduction channel, the basolateral currents within hair cells and the efferent actions on both hair cells and afferent nerve terminals.
- o Conduct molecular biological studies which isolate and characterize the molecular components of the transduction channel and associated regulatory proteins.
- o Identify neurotransmitters which mediate peripheral vestibular function .
- o Investigate metabolic requirements of the sensory organs and neurons in the inner ear and the mechanisms of production of inner-ear fluids and transduction elements (cristae and maculae).
- o Develop dynamic models that predict and explain patterns of afferent fiber activation on the basis of anatomical and biophysical properties of end-organs.

Reflex Control of Posture and Gaze

The goal is to understand the structure and function of neural circuits that transform vestibular sensory input into the motor output required for control of posture, locomotion, fine motor activities and gaze. Research is needed to:

- o Characterize the behavioral strategies and neural mechanisms used to determine posture and gaze during active and passive motion and during combinations of linear and angular acceleration in three-dimensions. Studies are also needed to define the effect of exposure to altered gravito-inertial fields on vestibular reflexes and their potential clinical consequences.
- o Characterize the anatomic and physiologic properties of neuronal pathways that constitute the vestibular system in animal models and normal humans . It is especially important to identify pathways involved in vestibular reflex control of neck, axial and limb muscles and to determine the signals they carry. Emphasis should also be given to understanding the neural substrates of otolithic organ reflexes and postural stabilization.
- o Correlate structure and cytochemistry of neurons in

different vestibular circuits with their response characteristics.

- o Characterize the biophysical, molecular and pharmacological properties of specific groups of relay neurons in postural and gaze reflex pathways.
- o Develop multidimensional models that explore the neural basis of dynamic and spatial transformations in vestibular reflexes.

Sensory Integration in Spatial Orientation, Perception and Motion Sickness

Understanding of the vestibular system requires knowledge of how vestibular signals interact with information from other senses to generate perceptions, movements and motion sickness. Research is needed to:

- o Describe sensorimotor and perceptual reactions to complex combinations of linear and angular accelerations and determine how vestibular, visual and proprioceptive inputs interact to generate perceptions of space and body motion.
- o Conduct anatomic and physiologic research to characterize the neural mechanisms that combine otolithic organ, semicircular canal, visual and somatosensory information to

generate perceptual and postural responses and that generate motion sickness.

- o Characterize the neural and humoral mechanisms associated with motion sickness and vertigo at the cellular and molecular levels.
- o Develop models that incorporate and test the understanding of the neural basis of postural and gaze control during active and passive motion.

Adaptive Changes in Vestibular Function

Studies of vestibular adaptation can both reveal basic principles of motor learning and lead to strategies for enhancing recovery from vestibular lesions. Research is needed to:

- o Determine at the behavioral level, both the extent to which the adaptive system can compensate for vestibular dysfunction and the sensory cues that are important in producing this compensation. Adaptive changes in both rapid and slow components of the VOR and in postural reflexes should be studied.
- o Conduct anatomic and physiologic research to determine where in the brain stem and cerebellum the neural changes responsible for adaptive alteration of vestibular reflexes occur and how these

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changes lead to the observed alterations in behavior.

- o Characterize the biophysical changes that occur at various neural sites during the adaptive process.
- o Analyze the pharmacologic and molecular bases of adaptive changes and the way in which they depend on expression of molecular mechanisms such as proto-oncogenes, second messengers or humoral factors.
- o Develop models that account for adaptive changes at both the biophysical and neural circuit levels.

Development and Aging of the Vestibular System

Knowledge in this area will both help us deal with developmental disorders and age-related declines in vestibular function and also contribute to basic understanding of vestibular mechanisms. Research is needed to:

- o Characterize at the behavioral level the progression of perceptual and reflex function during development and aging.
- o Conduct anatomic and physiologic research to determine the

mechanism(s) underlying development of central vestibular pathways, their relation to development of peripheral end-organs and the ontogenetic sequences of connectivity between vestibular neurons and motor and higher sensory centers in the brain.

- o Study how membrane and synaptic properties of hair cells and vestibular sensory and motor neurons change during development and aging and examine the roles of neural activity and neurotransmitters in producing those changes at the cellular level.
- o Determine the neurotransmitters present in vestibular brain stem nuclei over the course of development, maturation and aging.

Major Clinic Scientific Opportunities

There are six broad high priority areas for clinical vestibular research: prevalence and environmental factors; anatomic, physiologic and molecular bases; diagnostic methods and testing procedures; adaptive mechanisms; and medical and surgical therapy. These opportunities can be summarized as follows:

Prevalence and Environmental Factors

It is important to develop a better understanding of disease prevalence and possible associated environmental factors. Research is needed to:

- o Assess the distribution of balance disorders among different sectors of the population (including age, gender, genetic background and geography).
- o Identify environmental and occupational hazards that adversely affect balance.
- o Carry out a demographic study to evaluate the deleterious effects of prescription and over-the-counter medications which may produce central or peripheral vestibular alterations.

Anatomic, Physiologic and Molecular Bases

A better understanding of anatomic, physiologic and molecular bases of normal and abnormal balance processes is needed.

- o Determine modifications of neuronal vestibular pathways in animal and human pathologic specimens with known vestibular dysfunction.

- o Analyze the molecular and pharmacologic properties of specific groups of relay neurons in postural and gaze reflex pathways. The development of these properties from embryo to adult vestibular systems should be traced and changes that accompany aging examined.
- o Determine the coexistence of multiple neurotransmitters within single vestibular synapses.
- o Determine the excitatory and inhibitory neurotransmitters in the vestibular portion of the brain stem nuclei during development, maturation and aging.
- o Develop animal models to study the pathophysiology of vestibular diseases.
- o Examine structural changes in vestibular pathways in animal and human pathologic specimens with known vestibular dysfunction.
- o Investigate the molecular basis of acquired and congenital inner-ear disorders in humans and animal models.
- o Study and correlate peripheral and central pathologic abnormalities with biochemical and molecular changes in human subjects with well-studied disease.

Diagnostic Methods and Testing Procedures

Improved tests of balance function, a more standardized method of testing and research on pathology are needed.

- o Develop new tests to evaluate otolithic organ and vertical semicircular canal function in humans, including freely-moving subjects.
- o Develop tests to define the contributions of neck receptors to normal gaze and balance function and to vertigo and imbalance in pathologic conditions.
- o Develop new tests for vestibular function involving non-invasive recording of neural activity and/or reflex responses elicited by specific mechanical or electrical stimuli that activate vestibular afferents.
- o Develop new and improved methods for the evaluation of posture under static and dynamic conditions.
- o Develop standards for the more commonly-used diagnostic tests in the vestibular test battery.
- o Develop improved psychophysical methods for evaluation of vestibular function in health and disease.

- o Develop animal models to study the reliability and validity of new vestibular tests.
- o Develop methods to detect immunologic and autoimmune inner-ear disorders and neoplasms for the diagnosis of vestibular system disorders.
- o Validate currently accepted diagnoses or new diagnoses based on clinical pathologic correlation.
- o Conduct serial (longitudinal) studies of the changes in vestibular tests in patients with well-defined pathologic processes.

Adaptive Mechanisms

Adaptive mechanisms play an important role in normal balance function, compensation for disease and rehabilitation.

- o Analyze the molecular and pharmacologic bases of adaptive changes in the vestibular system.
- o Investigate the use of pharmacologic agents to modulate changes in adaptive compensation.
- o Extend studies of adaptive behavior to consider the conditions of unilateral and bilateral labyrinthectomy in an attempt to understand the response to vestibular damage.

- o Determine if there is a critical period for the initiation of vestibular exercises to facilitate recovery following the sudden onset of vestibular loss or dysfunction in humans.
- o Determine the optimal characteristics of vestibular stimulation necessary to facilitate recovery in different vestibular disorders.

Medical and Surgical Therapy

Medical and surgical therapy is frequently based on insufficient determination of efficacy. Standardized reporting of results is critical, and the efficacy of treatment should be determined.

- o Perform multi-center, prospective, controlled clinical trials which:
 - o evaluate medical therapy for vestibular symptoms.
 - o evaluate osmotic or renal loop diuretics for the treatment of episodic vertigo.
 - o compare medical to surgical ablative therapy in Meniere's disease.
- o Study the pathologic effect of vessel loop compression on the vestibular nerve and the effectiveness of vessel loop relocation.

- o Study the anatomic, physiologic and pharmacologic central nervous system correlates of neurectomy (partial and total) compared to labyrinthectomy in appropriate animal models.
- o Determine the effect of surgical manipulation of the cochlear nerve and the facial nerve intracranially and in the internal auditory canal.
- o Determine the effect of the peripheral and central molecular, biochemical and structural changes following various ablation procedures, e.g. labyrinthectomy and partial and total vestibular neurectomy.

Language and Language Impairments

Language is the uniquely human means of communication through which knowledge, belief and behavior can be explained and shared. The broad goal of research on language is to understand the nature of normal language function, including the underlying bases and mechanisms involved. A primary purpose of this work is to build the foundation necessary to develop and evaluate intervention strategies designed to improve and enhance communication

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for individuals with language disorders. The understanding of normal language (whether spoken, signed, or written) provides a basis for comparison in investigations of language disorders. It is critical to understand how language is produced and understood, what its biological and neural substrates and organizing principles are, how it is learned by children and how it is processed in adulthood.

Language acquisition takes place naturally for most children, including those who have normal hearing and those who are deaf with signing deaf parents. However, much research is still needed before normal language acquisition can be fully understood. There is an especially great need for data from all of the diverse groups of children that make up contemporary U.S. society. In light of the growing role played in society by different racial, ethnic and social groups, it is essential that these populations be properly represented in the subject pools for studies of normal language processes.

Individuals with normal hearing, as well as those with hearing impairment, may exhibit a disorder of language, that is, problems with language comprehension, production or use sufficient to interfere with interpersonal communication. In young children, these disorders frequently involve difficulty in the acquisition of the ambient spoken or signed language and may also lead to impairment in reading and writing. In adults and older children, persons with

language impairment include aphasic individuals who have lost their previous levels of language competence as a result of brain injury.

Disorders of language affect children and adults differently and pose different sorts of research questions. For the child who does not use language normally from birth or who develops the impairment in childhood, the disorder occurs in the context of a language system that is not fully developed. In contrast, impairment of language in adults disrupts a system that may be less malleable in the face of neural damage. As a result, although the broad goals of research on language disorders are similar whether the affected individuals are children or adults, the research agendas for these two groups are considered separately.

Both the review of research accomplishments and the discussion of research opportunities are divided into four general areas. The first of these concerns multicultural issues. This area receives emphasis because, with U.S. society becoming increasingly diverse, it is vital that evaluation of language ability be based on representative normative data. Moreover, there are indications that particular disorders may be more prevalent among certain groups; the development of preventive measures must rely on a clear understanding of the distribution of, and underlying bases for, language disorders in the population at large.

A second general area concerns language among deaf children and adults. Although much has been learned about the natural acquisition of signed languages, many details of the nature of this process are still unknown. Furthermore, many deaf children who acquire a signed language with little difficulty nevertheless have significant problems learning to read and write. Another important consideration is the fact that large numbers of individuals, including those whose hearing impairment was acquired during childhood, use spoken language as their primary mode of communication. The language acquisition and use of many of these people are characterized by difficulties not faced by most hearing persons.

A third general area deals with language disorders in children. These disorders can be discussed in terms of whether the language difficulties exist in isolation or in association with other problems and whether the factors interfering with language were present from birth or appeared later. Many children with isolated language problems present from birth are given the clinical label of "specifically language impaired". Children with language disorders occurring in conjunction with other developmental disorders include children with mental retardation and those with autism, among others. Finally, a number of children begin the language learning process normally but then acquire a

language disorder through infection, stroke or trauma.

The topic of language disorders in adults constitutes the fourth general area of concentration. The primary cause of such disorders is a focal brain lesion, caused by a stroke or head injury. Language disorders can also occur as a primary symptom of progressive dementing illnesses. In both cases, the previously-normal language system is changed in ways that can be devastating to the economic and social well-being of the affected individuals. Recent research on adult language disorders has emphasized the wide range and combination of language symptoms that can occur when the brain is damaged. This diversity of language impairments complicates efforts to understand the underlying functional bases for these disorders, to link particular symptoms to specific brain regions and to design effective rehabilitation techniques.

For all four of these areas of focus, current research opportunities emphasize studies that enhance understanding of the normal language system, while clarifying the various ways that language or language acquisition can break down. In all areas, successful intervention to address language impairments must be based on precise knowledge of the ways in which the disordered condition differs from the normal case.

Major Scientific Opportunities

Multicultural Issues

Specific research relating to multicultural issues should:

- o Include normative studies for different languages or social dialects which focus on the acquisition and use of phonological and grammatical forms as well as content and pragmatics. Investigations of the simultaneous or consecutive acquisition of two or more linguistic systems are also necessary. Such research can include investigations of "critical periods" for dual language learning and code switching.
- o Obtain basic epidemiologic information on the incidence and prevalence of and risk factors for speech, language and hearing disorders within African-American, Hispanic, Asian/Pacific Islander and Native American populations, as available data may not be generalizable across these groups.
- o Explore factors that might impede the efficacy of treatment, such as differences in cognitive or learning styles and cultural systems of belief.

- o Develop means of differentiating between language differences and disorders across cultural groups.
- o Investigate the incidence and prevalence of and risk factors for adult neurogenic impairment among culturally-diverse and bilingual populations

Language and Deaf People

Language Development in Deaf Children

Within this area, new studies are needed to:

- o Identify and describe patterns of acquisition of American Sign Language (ASL), particularly when access to a first natural language is delayed or incomplete.
- o Identify and describe patterns of language acquisition of children exposed primarily to systems or styles of signing in which the intention is to model or support English speech.
- o Identify and describe patterns of language acquisition among children exposed primarily to oral language.
- o Identify and describe patterns of language acquisition of children primarily exposed to spoken English through Cued Speech.

- o Identify and describe patterns of related cognitive, psychological and academic deficits accompanying delayed language acquisition.
- o Study the nature and value of language models provided by hearing adults in contact with deaf children. This research should include studies of the nature of receptive and productive aspects of English-like signing in addition to aspects of auditory, oral language models.
- o Identify deaf children who have language disorders by examining and describing the differences between normal and language-impaired deaf children.
- o Study approaches and techniques for the training of hearing parents of deaf children to sign or to facilitate the development of their children's spoken language, including the use of Cued Speech.
- o Identify and describe the processes of reorganization of the input language, particularly pidginization and creolization processes in language contact situations.
- o Identify and describe the invention of gestural language systems without formal sign input and the

relation of this process to eventual signed or spoken language development.

- o Study approaches and techniques for evaluating the potential of new technology and intervention strategies (such as auditory prostheses, laser disk technology, computer-based training systems) for promoting spoken language acquisition by deaf children.
- o Identify critical periods for both the natural acquisition and training of spoken or signed language by deaf children.
- o Develop the means of identifying language disorders in deaf children from different cultures.

Studies of Literacy in Deaf Children and Adults

Research should be conducted to:

- o Study the nature of processing text by successful deaf readers in an attempt to identify the most effective strategies, both auditory and visual, for the teaching of literacy to deaf people.
- o Develop and test new strategies and techniques for teaching English as a second language to deaf children who make use of their capabilities for visual language processing.

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- o Study the role of captioning, telecommunication devices, personal computers and other technological influences on the improvement of deaf children's literacy skills.
- o Study the relationships between speech processing and literacy in deaf children who use spoken language primarily.
- o Study the relationships among speech processing, sign processing and literacy in deaf children who use sign language.
- o Develop and evaluate appropriate and psychometrically-sound tests of psychosocial, intellectual and academic development and of first- and second-language acquisition by deaf children and adults under a variety of language use conditions.
- o Examine the interaction of acquisition of signed languages with spoken and written language and begin to characterize the resulting bilingualism. Studies of the order of acquisition of speech and signed language as they affect children's eventual acquisition of English language and literacy should also be conducted.

Basic Research on Sign Language Structure and Function

Investigators should be encouraged to:

- o Continue research on brain mapping for sign language functions.
- o Develop improved techniques for imaging sign language motions in three-dimensional space.
- o Conduct further investigations into how deaf people perceive and process visually. Studies could include investigations of the relation between limb control and vision and of the functioning of deaf people with visual disorders.
- o Conduct studies of the nature of parallel processing of language, such as the simultaneous processing of visual and auditory information.
- o Expand studies of the brain and language, such as those on the specialization of the cerebral hemispheres for language and other cognitive processing.
- o Study signed language and other languages used by the deaf community from functional/psychological and structural/linguistic perspectives. Studies should focus on the nature of the "creolization" of signed language and the effect of interaction of modality and

language structure, including contact signing.

- o Study signed languages other than ASL and varieties of signing within ASL, to identify the role of cultural and ethnic differences on signed language use by deaf people.
- o Study other signed languages, such as simultaneous communication and signed English, relative to the acquisition of English language by deaf children.
- o Relate infants' early acquisition of sign language phonology, assessed through tests of sign perception, to the acquisition of the higher levels of language, such as the acquisition of words (lexicon), word meanings (semantics) and grammar (syntax).
- o Investigate basic and higher level processes underlying vision, including the requirements of structured use of space and movement, the processing of complex dynamic arrays and the perception of motion and form.
- o Study the impairments to sign perception in deaf individuals who become aphasic or who sustain other cognitive impairments subsequent to brain damage.

Language and Its Disorders in Children

Bases of Language Disorders in Children

Research should be carried out to:

- o Examine the linguistic profiles of language-impaired children to determine if they vary according to the type of language being acquired (e.g., morphologically rich vs. sparse; flexible vs. rigid word order). The weaknesses observed in language-impaired children across these languages might reveal a common factor that may be the source of the disorder.
- o Examine the impact on language production of structural anomalies of the speech mechanism, such as craniofacial anomalies, long-standing tracheotomies or severe neuromotor impairment. The relative contribution of subtle motor speech problems to certain subtypes of language impairment is not known.
- o Study the normally-developing brain across the life span using a wide range of anatomic, physiologic and metabolic techniques. New advances in imaging the brain (anatomic, physiologic and metabolic) are particularly important to apply to studies investigating normal and abnormal development.

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- o Conduct multidisciplinary studies that combine imaging techniques with fine-grained behavioral analysis of processes underlying language development and disorders.
- o Determine the reason for the much higher prevalence of developmental language disorders in boys than girls.
- o Isolate those cognitive areas not represented in IQ tests and determine whether or not these areas interact with language functioning.
- o Determine to what extent language impairment may be secondary to psychosocial disorders.
- o Determine to what extent psychosocial disorders may result from a primary language impairment. There are special groups of children and youths who are at risk for language disorders and language-based learning disorders. These special groups include victims of abuse, neglect or drug- or alcohol-related lifestyles and homelessness and general victims of poverty.
- o Determine to what extent failures in communication and education are related to psychosocial factors.

Assessment

Initiatives are needed to:

- o Develop psychometrically-sound and culturally-fair and sensitive procedures for the measurement of language comprehension and production to be used in identifying and classifying language impairments.
- o Develop alternative assessment strategies including criterion-referenced testing, observational techniques, interview procedures and other informal strategies.
- o Explore the utility of state-of-the-art computer technology in language assessment including language sampling analysis.
- o Develop assessment strategies which have application, validity and reliability for identifying language disorders in children in a variety of assessment contexts (e.g., school, home, clinic).
- o Determine the most appropriate strategies for evaluating limited-English-proficient (LEP) children including the use of interpreters or informants.
- o Develop assessment tools for preschool populations, especially for children under three years of age.

Academic, Social and Vocational Impact

Research is needed to:

- o Study the relationship between language disorders and subsequent or concomitant learning disabilities.
- o Study the relationship in children between language disorders and social adjustment problems.

Intervention

Research must be conducted to:

- o Develop new intervention strategies based upon current theoretical models and establish their efficacy.
- o Determine the efficacy of currently available intervention strategies through both single subject studies and studies of children who clearly represent different types of language impairment.
- o Evaluate alternative models of service delivery such as family- or school-based programs versus direct delivery by speech-language pathologists.
- o Investigate the use of computers and recent technologic advances (for example, in speech recognition and speech production) in

intervention with language-impaired children and with deaf children learning oral language.

Language and Its Disorders in Adults

Brain-Language Relations

Detailed study is needed to:

- o Investigate the mechanisms by which recovery takes place and the basis for individual differences in recovery, using a variety of techniques for indexing brain activity in vivo (e.g., positron emission tomography, single photon emission computed tomography, event-related potentials).
- o Clarify the role of the non-dominant (typically right) cerebral hemisphere in the assumption of language functions following damage to the dominant hemisphere.
- o Relate specific symptoms to discrete brain regions using lesion analysis. A correlation between the breakdown of motor speech planning and structural brain lesions has been demonstrated, but other potentially localizable language skills (such as speech sound discrimination and oral word comprehension) are still inadequately mapped.

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- o Define by clinico-anatomic study the link between processes that are closely related to well-localized structures and those that are broadly distributed in the language zone.
- o Identify well-defined forms of selective impairments through detailed study of individual cases, including careful analysis of brain lesions and of functional language impairment.
- o Explore the role of various neurotransmitter systems in the mediation of particular language operations. The potential for pharmacological treatment follows directly from such understanding.

Analysis of Processes Underlying Language Disorders

Continued study is needed to:

- o Extend recent progress in the cognitive sciences to an understanding of the structure of normal language processing. For example, techniques involving real-time analysis of normal-language processes should be adapted to examine the nature of language pathology.
- o Distinguish the various forms of dysfunction that characterize oral/aural, written and signed

languages through comparative study of impairments in those modalities.

- o Elucidate the relationship between deficits that are specific to the language system and those that arise from perceptual, motor, or other cognitive disorders and clarify their relative contributions to complex language impairments. Information about such relationships could provide an important basis for the design of new treatments.
- o Continue comparative analyses of disorders in different languages, on the assumption that language-specific differences in symptoms can help reveal the linguistic and cognitive mechanisms that subserve language processing.
- o Develop artificial intelligence computer models that simulate normal language and its disorders.

Assessment, Intervention and Recovery

Specific opportunities in these areas include efforts to:

- o Demonstrate the efficacy and efficiency of the existing intervention strategies for aphasia treatment. Such demonstrations will require development of new approaches to the evaluation of

efficacy which use both single subject and group methods.

- o Revise and improve existing assessment procedures in order to bring them into line with current understanding of the relationship of patients' symptoms to causal underlying language deficits.
- o Develop a workable diagnostic scheme that provides a rational basis for the classification of patients and for comprehensive characterization of their deficits.
- o Develop functional assessment tools to evaluate the communicative abilities of language-impaired individuals.
- o Study the patterns of symptoms in patients with disorders in which deterioration instead of improvement is the natural course, for example, in Alzheimer's disease. Identification of predictors of the course of progression of language disorders in such patients would contribute to their management and provide a basis for approaching intervention.
- o Develop and evaluate computer-assisted instruction as an intervention strategy for working with aphasic individuals. In particular, strategies that capitalize on recent information concerning the functional basis of specific

language symptoms or that provide aphasic individuals with workable compensatory mechanisms should be developed.

- o Develop and evaluate alternative and augmentative communication systems for adults with language deficits. Adults severely-impaired in understanding and producing language may be capable of effective communication by alternative means, including computer-based systems.
- o Investigate social and cultural factors that have the potential to affect treatment outcome and evaluate the relative contribution of these factors to individual differences in response to treatment.

Comparative Language Studies

New initiatives are needed to:

- o Obtain normative data from healthy individuals of various ages to determine whether or not there are changes throughout the life span in the use of spoken, written or signed language.
- o Develop normative data on adults from a range of social strata and linguistic and cultural backgrounds to provide a basis for addressing the contributions of

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sociocultural factors, bilingualism and normal aging to language impairments.

- o Compare analysis of left- and right-brain damaged hearing subjects to determine the extent to which the right hemisphere may be involved in language use. Study brain-damaged users of sign language in order to understand hemispheric specialization for complex linguistic properties conveyed through spatial mechanisms.
- o Study the differences that exist between the language disorders of focally brain damaged patients and those that accompany more generalized brain dysfunction. Comparisons among the forms that

language breakdown can take secondary to these various causes have suggested important functional dissociations, but further study is required to clarify the nature of and underlying bases for these differences.

- o Develop normative data on the language functioning of diverse populations to provide a substantive basis for approaching their language disorders. The population of the United States is increasingly multilingual; as this population ages and its risk of stroke and consequent aphasia increases, there will be an increasing number of bilingual individuals with language disorders.

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BALANCE AND THE VESTIBULAR SYSTEM

Overview

The vestibular system maintains our balance and posture, regulates locomotion and other volitional movements and provides a conscious awareness of orientation in space and a visual fixation in motion. Disease, aging, exposure to unusual motion or altered gravitational environments (e.g., aerospace flight) can impair balance. In the United States, over 90 million people have experienced dizziness or a balance problem. For example, there are an estimated 38,000 new cases of Meniere's disease each year, one of many disorders that affects the vestibular system, and dizziness is the most common reason for seeking medical care in the over-75-year age group. A major consequence of vestibular disturbance is diminished capability and desire for purposeful activity. There may also be incapacitating effects on motor task performance and sensorimotor integration. Furthermore, motivation, concentration, memory, food intake and digestion and many other human activities may be adversely affected.

The vestibular receptors located in the labyrinth of the inner ear signal

motion and position of the head. Nerve fibers connecting these receptors to the brain provide information about head movements to produce vestibular reflexes. Brain circuits integrate vestibular information with other sensory signals from the eyes, skin, muscles and joints to produce a complex set of reflexes that interact to maintain posture and visual fixation in motion. In addition, the signals contribute to an internal representation or picture of body orientation and self motion.

The vestibular system is a complex, highly-interactive set of brain mechanisms capable of continuous adaptations to changes in the body and the environment. Because it has many components, there are many symptoms that indicate its malfunction, which range from mild discomfort to total incapacitation. Although balance is normally an automatic, unconscious process, it is essential for purposeful movement and effective communication.

Understanding how the vestibular system works is a scientific and technical challenge. The fluid-filled receptor end-organs that detect motion and head position are encased in bone and are difficult to access, manipulate and study.

The system is complex because it integrates many kinds of sensory information and utilizes a variety of neurochemical transmitters, and its malfunction results in a multitude of symptoms. The complexity of vestibular system function requires an interdisciplinary approach, including rigorous anatomical, physiological, biochemical, molecular biological, pharmacological, behavioral, perceptual and clinical studies. Biological data should be incorporated into mathematical computational models to examine and understand the integrated action of vestibular receptors and brain circuits in maintaining balance, orientation and the visual fixation and postural stability required for locomotion and other volitional movements. The resulting models should be utilized to predict changes that will result from pathologic conditions. Symptoms of these conditions should be evaluated in the context of the model predictions to reach a global understanding of the normal vestibular system and its pathophysiology.

Symptoms

The symptoms of vestibular system dysfunction are varied, reflecting the complexity of functional interconnections of the vestibular system with vision, tendon and joint position sensors, the digestive system and even the psyche (anxiety disorders).

Dysfunction of the vestibular system, particularly the inner ear and its interconnections with the brain, may cause hallucination of movement, variably described with such terms as dizziness, disorientation, vertigo, spinning, floating, rocking, lightheadedness, giddiness, sense of falling, imbalance, unsteadiness, or difficulty walking.

Symptom characteristics and the degree to which the patient is incapacitated depend upon the nature of the disorder. The symptoms may be episodic or continuous and may vary in frequency, severity and rapidity of onset. If there is damage to the system, the location (either in the inner ear or brain) as well as the magnitude of that damage play roles in the generation of symptoms.

Balance system dysfunction, if severe enough, can also provoke responses in the digestive system (nausea, vomiting and diarrhea), circulatory system (pallor, changes in blood pressure and pulse) and skin (perspiration, cold or clammy sensation).

The psychological impact of vestibular symptoms should not be underestimated. The fear of falling, with the chance of physical injury, adversely affects the individual's sense of independence and quality of life, particularly in the elderly. Similarly, the fear of a sudden attack of dizziness with socially-embarrassing ataxia, nausea and vomiting can cause individuals to become withdrawn. Subconscious fear may play

a role in agoraphobia and subtle dysfunction of the vestibular system may underlie difficulties in handwriting and reading. Vestibular dysfunction adversely affects the ability to perform the most routine activities, such as taking a shower, housekeeping duties and driving an automobile, and may prevent patients from holding high-risk jobs, e.g., telephone line-man, mechanic and bus driver.

Disorientation, i.e., wrongly perceived tilt or motion of the body relative to the environment, can arise from abnormalities in the vestibular system or result from subjecting the normal vestibular system to an abnormal environment.

Motion sickness is currently regarded as the brain's response to conflicting sensory messages about the body's orientation and state of motion. The sensors involved include the vestibular part of the inner ear, the eyes and body pressure and joint position sensors. Nausea, vomiting and poor concentration are symptoms provoked by "sensory mismatch."

Dizziness comprises a spectrum of disorders ranging from lightheadedness and giddiness to frank vertigo and dysequilibrium. Attempts to determine precisely the true incidence of vertigo/imbalance stemming from peripheral and central vestibular system dysfunction are foiled by the nature of the disorders and the complexity of the balance control mechanisms. However, it

is clear that balance disorders afflict a large proportion of the population, particularly the elderly.

Prevalence and Costs

Recent estimates exist for the prevalence of balance disorders in general, Meniere's disease and benign paroxysmal positional vertigo (BPPV). Examination of hospital statistics by discharge diagnosis shows that for those people under 65 years of age, 185,680 hospital days per year, on average, are provoked by dysequilibrium. In the general population (all ages), 347,000 hospital days are incurred because of "vertiginous syndromes," 202,000 because of "labyrinthitis" and 184,000 because of "labyrinthitis unspecified," with several thousands more accounted for by other balance disorders, e.g., Meniere's disease. The diagnosis of dizziness or dysequilibrium accounted for 221,000 of the primary discharge diagnoses in 1983. In the 45-to-64-year-old group, 1.3% of all visits to internists are related to vertigo or dizziness, affecting a relatively similar percentage of men and women.

Meniere's disease affects 15.3/100,000 individuals per year, resulting in about 38,250 new cases per year in the United States; in contrast, an incidence of 46/100,000 was found in a Scandinavian population, which would translate into 115,000 new cases per year in the United States. The prevalence figures are even more striking with a rate of 218.2/100,000. It is estimated there are

545,000 individuals in the United States with Meniere's disease.

In Japan, BPPV was determined to affect 10.7/100,000 individuals, with a great deal of inter-regional variability. The Japanese study determined that BPPV peaked in incidence in the fifth decade for both men and women, but that women were more commonly affected than men. BPPV occurred about two-thirds as frequently as Meniere's disease. The incidence of Meniere's disease in Japan is 16.0/100,000, very close to the incidence in the United States.

In looking at outpatient visits, 2.9% of visits to internists by the over-65-years-old group are because of dizziness or vertigo; in the over-75-years-old group, 3.8% of visits were for dizziness or vertigo, the number one reason for going to the physician. Seven percent of those in the over-85-years-old group present to the doctor because of dizziness or vertigo. These numbers reflect just the tip of the iceberg: an interview of those elderly at home showed 47% of the men and 60% of the women over the age of 70 complained of dizziness or vertigo.

In 1976, an estimated \$500 million was expended just on visits to physicians for dizziness or vertigo. Assuming, conservatively, a 5% rate of inflation, the costs for medical care alone will approximate one billion dollars in 1991. To this figure must be added the cost of diagnostic testing, as well as therapy. The cost of falls, which potentially lead to fractured limbs, hospitalization, surgery,

pneumonia and even death, must also be factored in. An estimated 15 to 23% of those over the age of 65 years who fall fall because of dizziness or vertigo.

The psychological toll of balance disorders on their victims is considerable; of those over the age of 65 years, at least 34% believe that dizziness keeps them from doing things they otherwise could do. This is not a happy way to spend one's "golden years."

If one considers military costs, the Army, Navy and the Air Force lose an average of 30 pilots and their aircraft every year to pilot disorientation and subsequent error. This loss of materiel conservatively represents an estimated cost of \$300 million per year. In times of strife, cost estimates are increased three- to four-fold. Also, two-thirds of all astronauts experience motion sickness that can severely reduce their effectiveness for the first three days into orbital flight.

Diagnosis and Treatment

Evaluation of vestibular function has improved in the last 15 years due to the development of tests based on modern anatomic and physiologic concepts and the application of technical advancements. For example, studies of individual neurons are providing data upon which models have been developed to understand precisely vestibular, visual and somatosensory inputs involved in gaze stabilization, balance and

orientation. Technical advancements in computer hardware have enabled clinicians to control angular and linear acceleration devices. Methods are being developed to measure different reflex functions (e.g., movements of the eyes, body or extremities) with the aid of microcomputers and programs that permit automatic analysis of data with accuracy and ease.

Tests cannot be limited to measurements of only one aspect of the balance system, however. Normal vestibular and balance function is a result of information about head movement and position from the inner ear, visuo-spatial relationships from the visual system and body relationships to the environment from the somatosensory system. Current methods of evaluating patients with vestibular disorders consist of a battery of tests that quantify the vestibulo-ocular reflex (VOR), the visual-oculomotor system and postural responses.

Better techniques to quantify vestibular function and its interaction with other systems, in addition to techniques that aid in early diagnosis such as magnetic resonance imaging (MRI) and techniques that permit evaluation of blood flow and metabolic function of the brain, are resulting in improved diagnosis and management of patients with vestibular disorders. Advancements in our understanding of the pathophysiology of vestibular disorders have also contributed to better medical and surgical management. For

example, mortality following resection of vestibular tumors has decreased from 50% to less than 1%.

Rehabilitation procedures are being developed that provide an additional approach to treatment for vestibular disorders. Specific vestibular exercise programs facilitate the rate and final level of recovery from vestibular disorders.

A Current View of Vestibular Function

Understanding the Molecular, Cellular and Neural Basis of Peripheral and Central Vestibular Function

Many of the vestibular reflexes serve to stabilize and coordinate movements of the eyes, head and body. These processes are collectively referred to as the vestibular reflex systems.

The vestibular receptor organs transduce the forces associated with head accelerations and changes in head position relative to gravity. The result is excitation of the nerves leading to control centers in the brain, which use these signals to develop a sense of orientation and to activate automatically muscles

that subserve movements of the eyes, locomotion and posture.

While the basic receptors and neural structures are genetically determined, the development and fine-tuning of vestibular function is a dynamic process that is dependent upon use and interaction with the environment throughout life. There must be an ongoing process of calibrating reflexes in order to adapt to physical changes in the musculoskeletal system during growth and development as well as to compensate for disorders (for example, inner-ear disease or visual loss) and to adapt to changes in the environment (such as an altered gravitational field, as in space). Thus, the proper responses to vestibular information require learning and memory.

Signal Transduction by Vestibular End-Organs

There are five vestibular end-organs. Two of these, the utricular and saccular maculae, transduce linear acceleration forces acting on the head, while the other three, the cristae of the semicircular canals, transduce angular acceleration forces. The five organs are so arranged in the temporal bone that they collectively provide the brain with a three-dimensional reconstruction of the position and motion of the head relative to the Earth.

Each end-organ consists of a sensory epithelium composed of sensory hair cells and supporting cells,

innervated by afferent nerve fibers from the vestibular nerve. Hair cells contain ciliary bundles (hairs) that protrude from their apical surfaces and are embedded in gelatinous structures, the otoconial membranes of the maculae and the cupulae of the cristae. These structures couple the forces acting on the head to the cilia resulting in bending of the cilia. Displacement of the ciliary bundles regulates the opening of transduction channels, thought to be located near the ciliary tips. When the channels are opened, an electrical transducer current flows into the hair cell and changes the voltage. The transducer current modulates ionic currents flowing through the basolateral surface of the hair cell. The basolateral currents alter the electrical response of the hair cell. In addition, one of the basolateral currents adjusts the intracellular concentration of calcium ions and thereby regulates the release of as yet unknown chemical neurotransmitters from the hair cell to the afferent fibers contacting (synapsing with) the hair cell. The result is a change in the nerve impulse frequency of the afferent fibers. Even at rest, hair cells continue to release neurotransmitters and thereby cause nerve fibers to have a background or spontaneous activity. Bending the cilia in one direction causes an increase in transmitter release and nerve activity; bending in the opposite direction results in a decrease. In addition to their afferent innervation, the receptor organs are also provided with efferent nerve fibers arising from the brain and contacting hair cells and afferent terminals. By means of this

efferent innervation, the brain can regulate ongoing sensory processing by the end-organs, as well as modulate their function on a longer time scale.

The afferent nerve fibers transmit their impulse activity to the vestibular nuclei and to the cerebellum. These structures are extensively interconnected with each other and with other centers in the brain stem and spinal cord that modulate the activity of the muscles of the eyes, head and body.

Reflex Control of Posture and Gaze

Afferent fibers from the semicircular canals and the otolith organs synapse with neurons in the vestibular nuclei of the brain stem, which form the starting point of neural circuits that regulate movements of the eyes, head and body. These movements take the form of reflexes that compensate for perturbations of head or body position.

Postural reflexes result from the integration of vestibular signals with input from visual and somatosensory receptors and other motor-control systems and generate patterns of activation of leg, trunk and neck muscles that stabilize posture and prevent falling. Maintenance of posture depends on the generation of different stabilizing movements depending upon the context in which adjustments must occur. Vestibular contributions to postural control are difficult to isolate from other postural stabilizing responses.

The direction of visual regard is regulated by gaze reflexes, which are better understood than postural reflexes. Such regulation is required to maintain stability of images on the retina, without which visual acuity is severely reduced. One such reflex, the vestibulo-ocular reflex (VOR), causes eye movement equal but opposite to head movement. When the head is rotated to the right, the horizontal semicircular canal in the right ear is activated and that in the left ear inhibited. Reflex circuits transform these changes in canal afferent activity into activation of muscles that move the eyes to the left by the same angle as the head movement so that the eyes remain directed to a fixed point in space as the head moves. In analogous fashion, when the head is displaced linearly upward (e.g. during running), saccular otoliths are stimulated; and by way of reflex pathways projecting to the ocular motor nuclei, the eyes are moved downwards to compensate for the head displacement.

Despite its seeming simplicity, the VOR requires complex and very accurate processing of semicircular canal, visual and otolith signals to generate the required compensatory eye movement. The angular acceleration signal provided by canals must be converted into a change in angular position of the eyes. Because the canals are not exactly aligned with eye muscles, signals from all canals must be combined to produce a spatial transformation from the three-dimensional coordinate frame of the canals to that of the eye muscles. These problems become even more complex in

the vestibulo-collic reflex, which must control over 30 neck muscles to stabilize the biomechanically complex head-neck system. Remarkably, the brain is able to perform these functions with relatively simple reflex circuits. The key is in having a parallel processing system with a multiplicity of neurons at each stage of the circuit, which gives the system great processing power and flexibility. Such systems can best be understood by computerized modeling techniques that can represent and explore their structure and computational capabilities.

The most direct neural circuits involved in gaze reflexes are three-neuron vestibulo-ocular and vestibulo-collic reflex arcs that interconnect vestibular afferent fibers, relay neurons in the vestibular nuclei and motor neurons that control eye or neck muscles. Each connection between a labyrinthine receptor and a pool of motor neurons involves several classes of relay neurons, each of which collects input from a different set of vestibular and visual or somatosensory receptors and projects to either a few related or many diverse motor pools. These classes of relay neurons may have distinct biophysical and pharmacological properties which allow them to be regulated independently and which may permit discrete therapeutic intervention to restore deficient reflex function once the mechanisms are known. In addition, the brain has a repertoire of more complex processes for regulating gaze which involve predictive switching, like that seen in postural systems. These are

implemented by more complex neural circuits and are as yet poorly understood.

Not surprisingly, to remain effective for visual stabilization, VORs must interact with the visual world during head movements. By receiving visual feedback during head movements, VORs may update their response properties to match what is required to maintain visual stability at any particular time and under a variety of circumstances. That is, the VORs exhibit adaptive plasticity. In addition, recent evidence has shown that VOR characteristics are affected by the vergence state and the direction of gaze in space, indicating that the VOR is not an isolated, simple reflex, but has response properties which are determined by integration of inputs from a variety of nervous system mechanisms and sensors other than vestibular ones.

Sensory Integration in Spatial Orientation, Perception and Motion Sickness

Spatial orientation is the relationship of the head and body to the Earth and to the Earth's gravity. The vestibular system and weight sensors provide information about how the body is oriented relative to the Earth, while other senses such as vision define the relationship of elements of the environment to the body. Balance and equilibrium are processes by which persons move and maintain head and body posture. Maintaining orientation and balance requires the integration of

information from many sources, especially the ears, eyes, skin, muscles and joints, and the transformation of these signals into coordinated patterns of muscle activity. To maintain balance and equilibrium under a variety of sensory and support surface conditions, combinations of sensory signals and muscle activity patterns must be interpreted appropriately.

Many sensory inputs are required for the production of a coordinated perception of head and body movement in space. The visual system tells the vestibular nuclei whether the reflexes produced by activation of vestibular end-organs provide the amount of gaze compensation that is required. Visual feedback is critical for proper operation of the VOR. This involves portions of the vestibulo-cerebellum, which play a role in adjusting the VOR when visual feedback indicates that it is not correctly stabilizing gaze. Disease of the cerebellum can result in faulty VOR calibration, poor gaze compensation during movement and imbalance and disorientation. Because vestibular signals interact with all of the major sensory systems, a large number of disease processes can impair balance and orientation. While it is clear that many brain structures are involved in determining orientation and balance, their precise roles remain to be determined.

The vestibular system operates largely automatically and without conscious awareness. When normal

coordination among the senses is disrupted by disease, a disturbing symptom, known as vertigo, occurs. Similarly, passive or unusual motion conditions (car and air travel, sea voyages and space flight) induce motion sickness, probably due to unusual combinations of signals from various sensors of motion. Mechanisms underlying the symptoms associated with vertigo and motion sickness are not well understood and are major unsolved problems as exemplified by the very high incidence of motion sickness which occurs during pilot training and reports that about two-thirds of the astronauts suffer from space motion sickness.

Gaze mechanisms involve not only eye movements but head movements as well. Frequently vestibular lesions that affect balance and orientation also affect the way the head is moved. This effect can lead to inappropriate stabilization of the head in an attempt to avoid unpleasant dizziness or vertigo, as well as possible dysfunction of the neck muscles and joints, which can also cause a sense of imbalance. Relatively little is known about the clinical effects of vestibular lesions on neck muscle and joint action or the reverse.

Adaptive Changes in Vestibular Function

A remarkable feature of the vestibular system is its potential for adaptive change (plasticity). This adaptive capacity allows the system to compensate for changes that occur

during development and aging or as a result of lesions. Most striking is the rebalancing of the system that occurs after loss of one vestibular nerve or labyrinth. The large postural imbalance and uncontrollable drift of the eyes that are seen immediately after a lesion disappear nearly completely over the course of a few weeks, although these signs can return if the system is stressed. To accomplish this recovery, the adaptive system must compensate for the loss of a large amount of ongoing neural activity that would normally come into the vestibular nuclei via the lesioned nerve. Experimental studies are beginning to reveal some of the neural and pharmacologic aspects of the multifactorial process that is involved.

Another example of adaptive vestibular control is the plasticity of the VOR, which is experienced by anyone who is fitted with a new set of prescription spectacles. Because the lenses magnify or reduce the size of the visual scene, images appear unstable when the head is moved: the VOR is still compensating for image motion that would be different without the lenses. This instability resolves in a few hours because the adaptive system changes the "gain" of the VOR and thus adjusts the speed of reflex eye movement appropriately for the new lens system. Experimentally, large changes in gain can be induced by having subjects wear magnifying and reducing lenses and even the direction of the VOR can be altered by appropriate pairing of head and image motion. Thus, in addition to its clinical

relevance, this VOR plasticity has become an important model system for understanding the neural basis of motor learning.

The role of the cerebellum in this process is the subject of current controversy. Cerebellar lesions abolish VOR plasticity, but one group of investigators believe that the cerebellum merely generates error signals that induce learning in the brain stem while another group has provided evidence that cerebellar circuits themselves can undergo plastic changes which account for VOR plasticity. This healthy controversy places the vestibular system at the forefront of attempts to understand sensorimotor systems. What is learned about the role of the cerebellum in the VOR will help scientists understand its role in regulating other types of movements.

As humans venture into space, they will be exposed for increasing periods of time to gravito-inertial fields different from those of Earth. Exposure to microgravity results in profound changes in the balance system. Extended exposure might be expected to have consequences on otolith reflexes involved with control of eye and head movements, posture and locomotion since those reflexes have evolved and function in the gravity of Earth. For example, the linear VOR, which depends on accurate information from otoliths, as well as reflexes dependent upon canal/otolith interactions, would be affected by exposure to space. Diminished accuracy

of eye movement control during head movement may result. Thus, in space, adaptive vestibular changes can lead to alterations in behavior and performance, as well as to changes in perception of spatial orientation. Such adaptive alterations can in turn produce difficulties when the individual returns to an Earth gravity environment.

Related Topics

The topics of development and aging, neural modeling and neurotransmitters and molecular biology relate to the four areas discussed in each section of this report, e.g., signal transduction by vestibular end-organs; reflex control of posture and gaze; sensory integration in spatial orientation, perception and motion sickness; and adaptive changes in vestibular function.

Development and Aging

The major goal of developmental studies of the vestibular system is to enhance the understanding of the cellular and molecular events leading to neuron proliferation, migration, differentiation and synapse formation. These activities are essential for neuron precursors to become vestibular neurons. In contrast, aging studies focus on postmaturational changes that may result in diminished function and/or cell death. Both types of studies are needed to gain a better understanding of how the brain processes vestibular signals and adapts to change.

One of the major challenges confronting developmental neurobiologists is to understand how connectivity and synaptic specificity are achieved. These issues are confounded in the vestibular system by the highly complex central vestibular network that relies on multisensory inputs for its normal activity and is composed of a highly interconnected network of neurons, with less topologic organization than in other sensory systems. However, there are advantages to the developmental approach which begins by looking at neurons and their connections when the basic components of their organization and function are simpler and more readily apparent. Examining progressively-older neurons and their more complex assemblies should result in further insights into how the system works. Studies of aging vestibular systems should enable us to determine whether the decline in vestibular function is a result of preprogramming (i.e., genetic influences) or the inability of the cells to sustain normal functional demands.

Following injury, the response of the adult vestibular system is characterized by a variety of responses which are species- and age-dependent. In lower vertebrates, hair cells in the inner ear and the vestibular nerve neurons have the capacity to regenerate. This capacity has not yet been demonstrated in mammals. Following the sectioning of amphibian axons, vestibular ganglion cells innervating individual receptor organs find their synaptic locus in the

vestibular nuclei. The new connections are functionally operative suggesting that their reestablishment and neuromodulatory function must be genetically imprinted. The central nervous system can compensate for deficits in vestibular function, and this capacity is greatest in primates, including man. Vision has been recognized as an important component in the compensation of VORs of primates and other mammals. There is a paucity of information, however, about the cellular and molecular mechanisms of the repair process. Such mechanisms are being recognized in new experimental work.

Neural Modeling

Because of the ability to measure precisely ocular motor and, to some extent, postural compensation in response to vestibular stimulation, mathematical modeling of these systems is an important method for relating their reflex behavior to the neural components that determine that behavior. Dynamic system models have contributed to defining the parameters that characterize the vestibular system. For example, the characterization of the dominant behavior of the VOR and certain postural responses in terms of time constants is a direct consequence of approximating the systems as first-order linear systems. Extensions of these concepts have led to defining parameters of VOR behavior in three dimensions and have given rise to model-based studies that relate these

parameters to fundamental organizational principles that determine the behavior of the VOR. Computational models and techniques that predict experimentally-obtained, behavioral results and uncover organizational principles that govern vestibular-induced behavior are needed.

Problems concerning integration of information from the vestibular end-organs and reflex pathways, as well as from other sensory systems, are important to study as a component of vestibular research of balance and posture, both in the civilian sector and in aerospace biomedical research. Computer modeling of neural network function is an important and indispensable tool for understanding how ensembles of neurons produce the required integration of information and generation of motor responses.

Theoretical neurocomputing studies also confront this key problem of information processing (sensory fusion) that has been solved by the brain in the vestibular system. The issue of information integration from different sensors is common to biomedical research, aerospace biology and neurocomputing research. Cooperation among fields will optimize success in each. This success can be achieved by identifying or creating research facilities that can integrate such multidisciplinary efforts.

Neurotransmitters and Molecular Biology

Transmission of vestibular signals from the ear to the brain is initiated when hair cells release molecules of neurotransmitters which react with receptors on the membrane of afferent nerve fibers. Several animal models have demonstrated central nervous system control of labyrinthine receptors by efferent fibers transmitting via acetylcholine. However, despite the identification of glutamate, histamine and other agents as potential afferent transmitters, conclusive evidence that any of these compounds serves as a primary neurotransmitter is still lacking.

It is important to characterize the nature of vestibular neurotransmitters and modulators and their interactions. Motor learning and motor control require continuous remodeling (adaptive plasticity) of neuronal connections and neurotransmission in central vestibular pathways. Understanding the molecular biology and pharmacology of those processes is an important future goal.

Understanding Pathophysiology

Our knowledge of the pathophysiology of many human vestibular disorders comes from experimental observations of the anatomy and physiology of the system together with careful clinical observations and pathologic examination

of human temporal bones and the central nervous system. The various mechanisms by which abnormal conditions may disturb the normal structure and function of the vestibular system can be categorized as: traumatic, inflammatory, degenerative, metabolic or neoplastic. These mechanisms may affect the end-organ and/or the vestibular nerve and brain as indicated below:

Trauma to the head may produce fractures or concussions which affect the inner ear or brain. Abnormal stimulation of the end-organ may be associated with otic capsule fistulization from bone resorption, change in the density of a cupula (cupulolithiasis) or an excessively long stapedectomy prosthesis.

Inflammatory destruction (labyrinthitis) is caused by bacterial or viral agents and immune disorders.

Degeneration of the end-organ may be produced by trauma, ototoxic antibiotics (aminoglycosides) and other drugs (loop diuretics, cis-platinum), vascular insufficiency (hemorrhages, infarction), hereditary disorders (Alport's syndrome, Waardenburg's syndrome) or surgical destruction (labyrinthectomy).

Metabolic disorders like diabetes mellitus, hypothyroidism, alcoholism or Paget's disease affect the vestibular system. Metabolic alteration (disturbed labyrinthine fluid composition or function) is commonly considered to be the mechanism by which Meniere's

disease and serous labyrinthitis affect the vestibular system.

Neoplastic alteration of the nerve fibers near the end-organ may be caused by an intra-labyrinthine schwannoma.

At the vestibular nerve level, trauma may be caused by mechanical compression resulting from arachnoid cysts, arterial loops and bone tumors. Inflammatory change is recognized as the pathologic alteration produced by viral and bacterial agents (vestibular neuronitis). Degenerative changes are associated with demyelinating disorders, such as multiple sclerosis and surgical transection. Neoplastic alteration is most commonly produced by benign tumors (vestibular schwannoma) but may also be associated with metastatic malignancies from other organs.

A variety of neurogenic disorders affect balance and different aspects of brain functions related to vestibular function, such as the control of gaze and posture. These include: hereditary and developmental disorders (congenital nystagmus, migraine, Chiari malformation); several familial ataxia syndromes involving different parts of the cerebellum; degenerative disorders (multiple sclerosis, progressive supranuclear palsies); various neoplastic lesions; vascular disorders, particularly of the brain stem and cerebellum (vascular malformation, infarctions and hemorrhages); and autoimmune disorders, including paraneoplastic syndromes.

Recent Accomplishments

Understanding the Molecular, Cellular and Neural Basis of Peripheral and Central Vestibular Function

Signal Transduction by Vestibular End-Organs

Recent technical advances in membrane biophysics have been applied to hair cells with the result that we now have a qualitative understanding of the events that link the bending of sensory hair bundles with the modulation of afferent impulse activity. Detailed studies in a few end-organs have shown how the various steps in the transduction process can be modified so that different hair cells and afferents become selectively tuned to certain aspects of the mechanical stimulus.

Afferent nerve fibers innervating a particular end-organ differ in their response properties. By the anatomic labeling of individual, physiologically-characterized fibers, it has been possible to relate this response diversity to the innervation patterns of the afferents, including their location in the sensory

epithelium and the types and number of hair cells that they contact.

Information has been obtained about the response of single vestibular afferent fibers to electrical activation of the efferent axons originating in the brain stem. Studies in alert animals have begun to suggest how efferent activity can modulate afferent discharge under physiological conditions. In addition to classical neurotransmitters such as acetylcholine, efferent neurons contain a variety of neuropeptides and transmitters (met-enkephalin and calcitonin gene-related peptide) that may influence receptor cells and afferent fibers in the end-organs.

Reflex Control of Posture and Gaze

Over the past five years considerable progress has been made in characterizing the sensory-to-motor transformations that occur in postural and gaze stabilizing reflexes:

- o The patterns of limb, trunk and neck muscle activation by postural reflexes have been described and the ways in which they vary with context are being explored.
- o Descriptions of the dynamic and three-dimensional spatial transformations that occur in vestibulo-ocular and vestibulo-collic reflexes have been obtained. The patterns of muscle activation that result have been described and models have been advanced that

successfully predict the muscle patterns that are used during the VOR and vestibulo-collic reflex.

- o Information has been obtained in humans and non-human primates on VORs and their interactions with vision in response to linear acceleration. Eye movement control has been shown to involve interactions among otolith, visual and vergence mechanisms, and further understanding of such control mechanisms should lead to the rationale for quantitative tools for clinical assessment of otolith function.
- o While understanding of the neuronal substrates of postural reflexes is at a rudimentary stage, great progress has been made in discovering the neural connections that mediate the transformations that occur in gaze-stabilizing reflexes.
- o The structure and function of vestibulo-ocular relay neurons that form the middle portion of three-neuron VOR arcs have been described in sufficient detail to reveal how convergent labyrinthine inputs to these neurons and their divergent projections to multiple motor pools could contribute to reflex transformations. Neural network models that explore these possibilities in a formal way are beginning to appear.

- o Beginnings have also been made in describing the more complex indirect pathways that contribute to the VOR and in determining how both direct and indirect pathways may be regulated by the cerebellum.
- o Research has begun to define the biophysics and pharmacology of neurons in vestibular reflex pathways.

Sensory Integration in Spatial Orientation, Perception and Motion Sickness

Originally, vestibular reflexes were conceived as being automatic and not subject to cognitive control. However, recent research has shown that we can use information from vestibular receptors during movement to determine where we started from and how far we have moved. We can also use the imagined location of near and far targets to adjust the magnitude of eye movements that compensate for linear or angular head movements. Thus, cognitive processing of vestibular information is not only important for determining spatial orientation but also for establishing proper control of VORs.

Great strides have been made in understanding the compensatory responses produced by the otolith organs during linear head movements by considering the eye movements that would be necessary to view real or imagined visual targets. Slower or faster

eye movements are required depending on whether targets are far or near. These compensatory eye movements even occur when targets are imagined in darkness.

Studies of this nature have revealed that the VOR is not a simple semicircular canal-mediated response. Instead it encompasses a broad range of reflex responses originating from each of the motion sensors in the labyrinth. Inputs from these sensors are directed toward the vestibular nuclei, where they are integrated with information from the visual and somatosensory systems and from proprioceptors to form signals that direct gaze movements and stabilize posture and equilibrium. Cerebellar participation is essential for proper operation of these vestibular reflexes.

Models of the VOR and of vestibular compensatory eye movements have shown that there are at least two components to the reflex. One operates rapidly to produce quick changes in eye position and eye movement in response to head movement. The other has more sluggish characteristics but outlasts impulses of acceleration and is largely responsible for the characteristics of eye movements induced by rotation. This component has recently been shown to be oriented around gravity. Both components are subject to adaptation, and these types of motor learning are controlled by various portions of the vestibulo-cerebellum. Lesions of the cerebellum cause deficits in compensatory movements, as well as disorientation and dysequilibrium.

The current theory of motion sickness holds that it is a by-product of the process of adaptation to unusual sensory inputs or motion conditions. It has been demonstrated that subjects have a remarkable ability to adapt to unusual motion conditions, although the process of adaptation is often associated with motion sickness. When adaptation is finally achieved, the vertigo and/or motion sickness disappears.

Adaptive Changes in Vestibular Function

As described above, the vestibular and balance control systems have the remarkable ability to maintain useful function in many novel motion environments and to adapt to abnormal function of one or more of their components. The ability of these systems to adapt systematically sensory, neural and motor components in order to achieve useful compensation must be more thoroughly understood. Advances in vestibular health care, particularly rehabilitation, should be developed from sound scientific concepts. Therefore, the significance of central nervous system phenomena associated with the adaptive process (such as neural sprouting, reactive synaptogenesis and long-term potentiation or depression of synaptic transmission), which are thought to play important roles in adaptive control, must be measured and correlated systematically with behavior. Some accomplishments that have opened

exciting new areas of investigation include:

- o The essential role of adaptive plasticity in normal function of the vestibular system has been recognized. This system is unusual in that it is not static. It is constantly changing in response to changes in the environment or internal elements (for example, inner-ear or central nervous system diseases).
- o Neural pathways that may play a role in adaptive changes have been identified. As indicated above, there are several possible models of the neural basis of VOR plasticity. Information is also beginning to emerge about the neural changes that accompany recovery from unilateral labyrinthectomy.
- o The pharmacologic aspects of adaptive changes and their roles in transmitter synthesis as well as the release, number and sensitivity of receptors have been described.

Research from both U.S. and Soviet space flights has shown alterations in eye movement and postural control, as well as in perception of spatial orientation, both during flight and after return to Earth. It appears that the plasticity of vestibular reflexes, for the most part, allows adaptation during short duration missions, but the effects of vestibular system performance after long exposures is unknown. The precise characteristics

of adaptive plasticity in this unique environment and the vestibular function involved in readaptation to Earth's gravity after long-duration exposure to space remain unknown. Vestibular and balance system adaptation to altered gravito-inertial environments has not been measured systematically.

Related Topics

Development and Aging

Recent work suggests that a synchrony may exist between structural maturation of the vestibular end-organs and central vestibular nuclei. In recent developmental studies of the vestibular system, classical approaches have been supplemented with molecular biological probes and innovative techniques. For example, connectivity of the vestibulo-ocular and vestibulo-spinal pathways has been studied by the application of fluorescent dyes (e.g. Lucifer yellow, carbocyanine) in brain slices and on cultured brain stem and spinal cord preparations; immunocytochemical probes are used to examine the vestibular pathways and the maturation of otolith organs; and brain slice preparations are employed to obtain membrane and synaptic properties of developing vestibular sensory neurons.

From the combination of electron microscopic methods and tract tracing techniques, we are beginning to determine the steps in the assembly of synaptic inputs to the vestibular brain stem nuclei. Specifically, the first

synapses formed in the chick's lateral vestibular nucleus are not formed by primary vestibular fibers but appear to be derived from fibers of central origins. It is important to determine the steps in the assembly of various synaptic inputs to vestibular neurons in order to test what role synaptic formation may play in orchestrating developmental events.

The aging vestibular system also shows interesting changes in the structure of sensory and non-sensory components. Senescence has long been associated with increased vestibular dysfunction in older subjects who have more difficulty with vestibular test performance than younger subjects. Animal studies indicate that neural vestibular components in the ear and brain tend to accumulate a so-called aging pigment or lipofuscin. This and related substances, accumulated during the process of aging, may place a burden on the normal functioning of neurons and may diminish their response to functional demands. The characteristics of structural, functional and behavioral modifications of the aging vestibular system need to be studied. We need to determine if diminished efficiency with age is due to irreversible preprogrammed cessation of function or to reversible cellular modifications.

Neural Modeling

In the past decade, there has been increasing interest in mathematical modeling of vestibular systems at the level of the end-organ, neural pathways

and reflex behavior. Models are important because they formalize concepts, organize data and predict responses. Modeling reflex behavior by applying control system analysis, as developed and used in electrical engineering, has been augmented by developing mathematical descriptions of the functioning of parallel neural networks. This conceptual advance is important because it shows that it is possible to reproduce neural behavior by layers of interactive adapting elements. The vestibular system is a natural entry point for the use of such models linking biological and theoretical neural network approaches.

It is not sufficient merely to set up neural networks with a multitude of parameters and adapt them to fit arbitrary data. Studies need to be conducted to derive learning algorithms from fundamental principles that are consistent with mathematically-formalized, behavioral constraints. Using realistic models of cellular dynamics as derived from pharmacodynamics could also be helpful in uncovering the principles that govern behavioral learning in the vestibular system such as adaptation and habituation.

Neurotransmitters and Molecular Biology

Considerable progress has been made in identifying the neurotransmitters involved in the vestibular system. The field of potential primary hair cell neurotransmitters has

been narrowed to glutamate-related excitatory amino acids. The transmitters liberated from the eighth nerve fibers onto brain stem neurons have not yet been specified, but the inhibitory transmitters of the vestibular commissural systems include gamma-amino-butyric acid and glycine. Since data from recent studies on other systems indicate that several neurotransmitters may coexist in the same synapse, this possibility should be investigated in the vestibular system.

Advances in Diagnosis

Thoughtful history taking based on a knowledge of the anatomy, physiology and pathology of the vestibular system provides the core of diagnostic information necessary for clinical practice. This information is enhanced by tests which describe the integrity of vestibular reflexes.

Testing - Physical

Advances in modern technology have made the evaluation of vestibular patients a quantitative science. Because the VOR is the best understood vestibular reflex, scientists have taken advantage of the reliability of eye movement measurement to develop a battery of vestibular function tests to study the integrity of the various VORs.

The function of the cristae of the horizontal semicircular canals, sensors for angular acceleration, can be

measured with the caloric test by irrigating, under precisely controlled conditions, the external ear canal with water at different temperatures. The resulting eye movements can be accurately and efficiently measured with the aid of small laboratory computers.

Other methods of evaluating semicircular canal function include the use of rotary platforms and measurements of VOR responses to obtain precise information about the relationship between stimuli and reflex eye movements.

The function of the linear acceleration sensors of the ear (the otolithic organs) has been measured by observing ocular reflex responses to stimulation with various devices including the parallel swing. Also, the evaluation of balance has improved greatly with the use of platforms which measure movement of the patient's center of mass with sensitive force transducers whose signals are analyzed by computer.

The contribution of the visual system to orientation is evaluated by measurement of visual oculomotor reflexes, including the use of tests for smooth pursuit function and optokinetic nystagmus. These tests evaluate the ability of patients to follow small visual images (the smooth pursuit system) or the movement of large visual scenery (optokinetic function). The most revealing information is obtained by combining visual with vestibular stimulation as described above. Given

the extent of neural pathways subserving vestibulo-ocular and visuo-oculomotor reflexes, information from these tests provide methods to evaluate large areas of the brain. The combination of such tests in the modern vestibular test battery has allowed the identification of the location within inner ear or specific brain regions of vestibular lesions, a quantitative estimate of the magnitude of the deficit and the ability to follow the course of the disease. Useful features of these tests are their non-invasive nature, their ease of administration and minimal discomfort for the patient.

In addition to these standard tests, new tests are being developed in animal studies and being investigated in humans. Several of the new human vestibular tests are designed to evaluate anterior and posterior semicircular canal function (in addition to horizontal canal function) by rotating subjects about different axes to optimize stimulation of the specific canals. A problem associated with attempts to activate vertical canals is the simultaneous stimulation of the otolithic organs and the complexity of measuring the resulting eye movement. New methods, such as video-based monitors, are being developed to deal with the latter problem.

Of all the available vestibular tests, only the caloric test allows specific location of a lesion in one of the ears. All other tests using physiologic stimuli represent the response of a multiplicity of nerve centers. In recent years, however, animal experiments have provided

information that suggests the possibility of evaluating lesions specifically located in the vestibular nerve. Vestibular-evoked potentials have been obtained from the nerves of the semicircular and otolith organs in several animal species. Responses from direct electrical stimulation of the vestibular nerve also have been obtained in animal and human experiments.

A major deficiency is the lack of tests for otolithic and proprioceptive reflexes. Tests are being developed to evaluate these systems under operational conditions in freely-moving animals and human subjects. There is interest in the evaluation of head and eye interactions in humans with normal and abnormal vestibular function. Also lacking are tests for the quantitative evaluation of the subjective sensation of motion and of dizziness in patients.

Also, there are no tests that allow the physician to assess the possible role of neck muscle or bone lesions in the production of dizziness, vertigo or postural imbalance.

Testing - Laboratory/Radiology

Tests for antibodies to inner-ear antigens have recently been developed and appear to help in the diagnosis of inner ear autoimmune disease.

Fine resolution imaging has improved with the development of better software and technology for both computed tomography (CT) and

magnetic resonance (MR) scanning. Improved use of contrast and speed of image acquisition have made it possible to detect smaller lesions and permit dynamic CT/MR scanning to evaluate blood flow in small regions of the central nervous system. The development of techniques for imaging the individual receptors in the labyrinth would improve diagnosis and understanding of the disease process.

Advances in Treatment

Medical Therapy

Advances in basic research have identified probable transmitters and receptor sites in the vestibular periphery and central connections. These findings have not only led to clinical trials of transmitter agonists and antagonists but have also resulted in the realization that most pharmacologic agents with potential central nervous system activity may interfere with normal vestibular function. For example, long-acting sleeping pills and tranquilizers are now recognized as a major cause of injuries from falling in nursing home patients.

Clinical trials of antihistamines, anticholinergics and phenothiazines with improved experimental design have been conducted, and these agents appear to be helpful in relieving the symptoms of vertigo. Unfortunately, these agents have side effects such as drowsiness and dry mouth and eyes.

The reduction of symptoms, presumably related to increased endolymph fluid, with systemic diuretics appears to be helpful, at least in the early stages of Meniere's disease.

Further studies using systemic, transtympanic or intralabyrinthine aminoglycosides show good results in reducing vertiginous attacks in patients with Meniere's disease but have the potential side effects of loss of hearing or severe loss of vestibular function and oscillopsia (a condition in which objects appear to move up and down or from side to side).

The recognition of the relationship between associated disorders such as depression, anxiety attacks and panic attacks and vestibular disorders suggests the need to investigate these disorders as well. Recent advances in the pharmacologic control of these disorders is encouraging.

Surgical Therapy

Surgical treatment strategies in peripheral vestibular disorders may be grouped into ablative and non-ablative protocols. Ablation procedures relieve the balance symptoms by denervating the altered labyrinth without correcting the lesion. These procedures are 90 to 99 percent effective in relieving episodic dizziness provided that the ablation is complete. If hearing is not useful in the unilaterally-diseased ear, labyrinthectomy (transcanal or transmastoid approach), is a short,

relatively-safe technique. If there is useful hearing in an unilaterally-affected ear, selective vestibular neurectomy, via a middle cranial fossa (MF) or posterior cranial fossa (PF) approach, is now the accepted procedure. The MF approach is technically more difficult and is associated with a higher incidence (25 percent) of temporary facial nerve weakness than the PF approach. However, it has a higher success rate (95 to 98 percent) for vertigo control than the PF method (80 to 90 percent) because of the natural separation of the vestibular and auditory nerves in the internal auditory canal. The separation of these two groups of fibers in the posterior fossa section of the eighth nerve is arbitrarily created by the surgeon.

Selective transection of the posterior ampullary (singular) nerve has been successful in more than 90 percent of patients with chronic disabling positional vertigo (cupulolithiasis). Associated sensorineural hearing loss has occurred in fewer than five percent of the cases.

Non-ablative procedures correct the pathologic derangement responsible for altered vestibular function. Several examples of these are: removal of chronic inflammatory tissue (cholesteatoma) from a bony semicircular canal fistula, removal of an excessively long stapedectomy prosthesis, removal of chronic inflammatory tissue from the oval and round windows and repair of a perilymphatic fistula in the oval or round

windows caused by direct or indirect trauma.

The efficacy of vessel loop relocation from the intracranial portion of the eighth nerve has been variable and remains undetermined at this time.

Rehabilitation

The use of exercises to improve gaze and postural stability in patients with vestibular disorders has become increasingly popular in the past five years. The exercises are based in part on anecdotal evidence of improved function in patients following exercise intervention. The exercises are also based on the results of animal research demonstrating that visuo-motor experience can increase the rate of recovery following unilateral vestibular loss and that preventing visual inputs or movement can delay the onset of recovery. Research on human subjects is in progress, but the results are not yet known.

Outcome measures of treatment efficacy include many of the standard methods for quantifying balance but are often dependent on subjective reports. A questionnaire that measures the patient's perception of the extent to which the vestibular or balance disorder is handicapping has been developed and validated.

Program Goals

There is a need to expand the knowledge of normal and abnormal structure and function of the vestibular system using recently developed techniques in a broad spectrum of scientific disciplines. Scientists must be trained and research support provided to establish a knowledge base commensurate with other fields of medicine. In order to achieve this goal there must be increased study of both human pathologic material and animal studies.

Understanding the Molecular, Cellular and Neural Basis of Pheripheral and Central Vestibular Function

Sensorimotor systems such as the vestibular system depend upon the integration of information from many sensory sources which must be transformed to control many different muscle groups, thus posing a unique experimental challenge. Vestibular functions, especially those involved in stabilizing gaze, are sufficiently simple and stereotyped in their action to raise

the hope that vestibular research will yield the first breakthrough in understanding a complete sensorimotor system and major advances in understanding clinical problems. To achieve this goal a broad array of modern molecular, pharmacological, biophysical, anatomical, physiological, psychophysical and kinesiological techniques need to be mobilized to study the system at the subcellular, biophysical, neural circuit, perceptual and behavioral levels. Data obtained from these studies must be integrated into detailed computational models that will permit the knowledge of the system to be tested and used to predict outcomes of new basic and clinical experiments.

Signal Transduction by Vestibular End-Organs

At the level of the vestibular end-organs, the overall goal is to understand the response of vestibular nerve fibers in terms of cellular mechanisms, which in turn must be related to cupular and otolithic membrane dynamics. This understanding will require detailed studies of the sensory epithelium, including hair cells and their afferent and efferent innervation. Since afferent nerve fibers which innervate different parts of an end-organ vary in their properties, it will be important to study mechanisms on a regional basis. The following goals have been identified:

- o Use advanced imaging techniques to study the mechanics of the cupulae and otolithic membranes

and the hair bundle attachments to them. Relate the mechanical properties to the physical and molecular properties of these structures.

- o Relate the biophysical properties of the transduction channel to the ultrastructure of the hair bundle and to biochemical events taking place in the cilia. Develop the biochemical techniques needed to isolate and characterize the molecular components of the transduction channel and the associated regulatory proteins.
- o Characterize the biophysics and cellular biology of hair cells and afferent terminals, relating them to their location in the sensory epithelium and to the structure of the hair cells (Type I or Type II).
- o Study the pharmacology and biochemistry of afferent and efferent neurotransmitters. Use recently developed techniques of chemical neuroanatomy to determine if there are several populations of afferent or efferent neurons which are distinguishable on the basis of their transmitter biochemistry.
- o Define the role of efferent neurons, including an analysis of the effects of efferent activation on receptors and afferent synapses. Afferent-efferent interactions need to be understood in terms of electrical,

- chemical, metabolic and neuromodulatory mechanisms. Use alert animals to determine the behavioral conditions that lead to changes in efferent activity, to characterize the profile of the resulting changes and to investigate their influence on the end-organ.
 - o Develop experimental models for peripheral vestibular dysfunction, taking advantage of species differences and genetic variations.
 - o Characterize the mechanisms regulating fluid, electrolytes and metabolites of the inner ear.
- Reflex Control of Posture and Gaze***
- There is a need for further understanding of the reflexes associated with posture and gaze at the neural circuit level, including the basic reflex connections and the more complex pathways that supplement and modulate their actions. The overall goal is to mount a comprehensive, interdisciplinary initiative to understand the function of these critical reflexes. Some specific goals are:
- o Undertake a comprehensive analysis of the neuron populations that participate in gaze-stabilizing reflexes in order to correlate their biophysical and pharmacological properties with their structure and function.
 - o Identify with anatomic and physiologic techniques the neural substrates of vestibular reflexes acting on neck, axial and limb musculature.
 - o Develop computational models that will foster understanding of how populations of neurons can implement postural and gaze reflexes.
 - o Determine at the behavioral level, which strategies the central nervous system uses to control movement and posture. Define the relative roles played by sensory inputs from semicircular canals, otolithic organs, neck muscle proprioceptors and the eyes in determining the dynamic and spatial characteristics of movement.
 - o Determine the discrete or continuous nature of sensory-to-motor transformations in postural reflexes of animal and human subjects. Undertake experiments to record neural activity related to these reflexes in animals.
 - o Define the mechanisms underlying otolithic organ control of eye movements and how they are integrated with visual mechanisms, particularly during linear accelerations that occur during normal behaviors, such as locomotion and postural sway. Determine how otolithic organ and

semicircular canal control of eye movements interact during the complex motions that include components of linear and angular motion.

- o Develop tools for the quantitative assessment of otolithic organs' function in normal persons and in those with clinical indications of vestibular malfunction.

Sensory Integration in Spatial Orientation, Perception and Motion Sickness

Natural Motion Versus Passive Motion

The role of the VOR is to stabilize gaze so that images can be held on the retina and seen. To date, most studies of the VOR have been performed by rotating subjects with their heads and bodies fixed. However, the vestibular system works in freely-moving individuals who are changing both head and eye positions. Although it is technically difficult, it is essential that we address the problem of gaze stabilization in freely-moving subjects to gain a full appreciation of the range and capability of the vestibular system. Only when we study the vestibular system in its natural state will we begin to understand the nature of the deficits that occur after unilateral and bilateral peripheral vestibular disease and central vestibular or cerebellar disease or in aging. These studies should be conducted in animal and human subjects who are engaged in

full-body movement while their eye and head movements and single-unit activity from the brain stem are monitored. Reactions produced by controlled-passive head and body movement should be compared with reactions in freely-moving subjects.

Processing and Modeling of the Vestibulo-Spinal System

A particularly important aspect of otolithic-vestibular, nuclear and vestibulo-cerebellar processing is that information about upright stance and balance is created from otolithic organs and other signals that provide a sense of the upright and from the feedback information from joint and muscle proprioceptors. It is critical to have a better understanding of the various parts of the vestibulo-spinal system. At least part of the problem is that there has been little attempt to model the vestibulo-spinal system other than to predict muscles that would be activated by individual semicircular canals during natural head movements. We need dynamic models that match the characteristics of the output signals to the characteristics of the body part upon which the vestibular signals operate.

Neural Basis for Spatial Orientation

Orientation in space is a complex function that involves the vestibular system as well as cerebral structures and cognition. The neural basis for spatial orientation is poorly understood, and

promising leads obtained from blood flow studies, single-unit studies and the results of specific lesions should be pursued to enlarge our conceptual models of this important area.

Motion Sickness

Central nervous system changes in cellular and hormonal activity associated with motion sickness must be characterized in appropriate animal models and humans.

Adaptive Changes in Vestibular Function

If we are to harness the capabilities of the adaptive system to optimize vestibular function at all ages and to restore function after peripheral or central lesions, we must more fully understand its operation at the behavioral and neural levels.

At the behavioral level we need to:

- o Develop an understanding of the physical stimuli that govern adaptive behavior. For instance, evaluate the relative roles of inputs from central versus peripheral retinal receptors, inputs from various vestibular end-organs and inputs created by active versus passive motion.
- o Understand what aspects of such vestibular performances as reflex size or timing, perception of space or motion and regulation of static

and dynamic posture can be altered by the adaptive system.

- o Investigate the ability of the vestibular system to adapt to special environments, such as under water, high performance aircraft and the various conditions of space flight.

At the neural level, we must define the anatomical, physiological and biochemical changes in specific neural circuits that underlie adaptive behavior and develop models for systematic investigation of how such neural changes can contribute to compensation for functional deficits in the vestibular system. Three sequential goals for achieving this are:

- o Determine at which site(s) the neural changes responsible for adaptation occur using anatomic, histologic and physiologic techniques.
- o Analyze the biophysical, pharmacological and molecular bases of these changes and evaluate the possibility of manipulating them therapeutically.
- o Develop neural network models which determine whether the observed neural changes are sufficient to explain adaptive vestibular performance and to predict the effects of various therapeutic interventions on adaptive performance.

In addition, further research is necessary to understand the cellular source of the slow and rapid components of the VOR, how the different areas of the vestibulo-cerebellum are interrelated and the cellular mechanisms of their action. An understanding of the motor learning that is involved in control of both the rapid and slow components of the VOR is likely to involve an understanding of how the mossy fiber input and climbing fiber systems originating in the inferior olive interact with Purkinje cells. Only when these interactive loops are understood will we be able to model and predict the adaptive capability of individuals to alter their responses to changing visual and vestibular inputs, including altered gravitational environments and after unilateral and bilateral disease of the labyrinth.

Model validation must proceed at all levels (neural, sensory, motor, perceptual and behavioral). Modeling is an excellent way to organize our thinking about the dynamics of the interactions we seek to understand. New and potentially more powerful approaches to mathematical modeling are to be encouraged along with continued reexamination of existing models in the light of new experimental data.

Related Topics

Development and Aging

A central issue in developmental neurobiology is the interdependence of

axons and the neurons they innervate and how this relationship is influenced by growth, experience, injury and aging. To understand assembly of the vestibular system, systematic studies are required to define the sequence of innervation and synapse formation on central vestibular neurons. Specific issues are:

- o Study the onset of signal processing in the vestibular system during development and the correspondence between peripheral and central structural changes leading to the onset of vestibular function.
- o Determine the role of neurotransmitters and neural activity in vestibular neuron development (at the level of identified cells).
- o Study the emerging membrane properties of identified vestibular neurons.
- o Study the responses of developing vestibular neurons to vestibular and non-vestibular stimulations. Studies should be performed on both normal developing neurons and those in partially-deafferented brains.
- o Compare the morphogenetic and electrophysical relations between vestibular axons and Type I and Type II hair cells in the end-organ and examine how peripheral structure changes during

synaptogenesis in the developing labyrinth.

- o Compare central nervous system and peripheral nervous system synaptic interactions to determine what factors influence development at different ages, focusing on critical periods of development.**
- o Determine the onset and progression of sensory integration of inputs from specific vestibular end-organs to vestibular neurons in the brain stem. This time course should be related to the functional changes in vestibulo-ocular and vestibulo-spinal reflexes as a function of age.**
- o Determine the behavioral changes associated with the aging process and design strategies for preventive and rehabilitative intervention.**
- o Determine the anatomical and biochemical changes associated with aging at various parts of the reflex. This study should include the quantification of cellular changes in the receptor organs (sensory and supporting structures), primary afferents, vestibular nucleus neurons and motor efferent elements.**
- o Study the differential role of aging in the various cellular elements of the vestibular reflex to find familial and idiopathic causes of premature**

aging in the inner ears and central vestibular centers.

- o Use animal models, including mutational variations, to elucidate the causes and course of the aging process.**

Neural Modeling

Impressive strides have been made in understanding areas of the vestibular system where it has been possible to model the neural input and output, thereby formalizing the transfer functions that are performed by the central nervous system. There is a continuing need to model the VOR and perceptual-motor responses to various combinations of semicircular canal and otolithic organ stimulation. In addition to animal models and human pathologic material, concentrated efforts must be made to develop mathematical and computer models to complement and supplement ongoing experiments.

Wherever possible, strategies that the central nervous system uses to control movement and posture should be defined using mathematical and computational paradigms and models at the behavioral level, as well as at the level of neuronal networks and single neurons. Specific goals are to:

- o Develop mathematical and computer models for systematic integration of the knowledge base on vestibular function and adaptation into a formal**

understanding of the function of vestibular neural networks.

- o Develop cooperative studies to investigate vestibular and balance problems in special environments, such as underwater, aviation and aerospace. Incorporate mathematical-computer modeling of vestibular systems with the goal of integrating experimental knowledge into a theoretical understanding of the brain as a neurocomputer.
- o Seek experimental verification of models under simple as well as complex conditions of motion.

Neurotransmitters and Molecular Biology

The molecular properties of neurotransmitters of the vestibular system should be investigated. Multidisciplinary approaches should be combined in the same animals, whenever possible, in order to delineate cause and effect relationships. Use should be made of animal and human material to study the presence of neurotransmitters in adult subjects as well as during development. Modern techniques for neurotransmitter identification (e.g., in-situ hybridization) and immunochemistry should be combined to elucidate the synaptic processes and their development. Specific questions requiring investigation are:

- o Characterize molecules released in response to electrical and chemical stimulation with pharmacologic and molecular techniques.
- o Examine molecular changes that accompany documented and measurable deficits in vitro and in animal models to determine their relationship to the deficits, causal or not, and their mechanism of action.
- o Document coexistence of multiple molecules with neurotransmitter and neuromodulator properties in individual neurons. Their interactions should be investigated.
- o Perform experiments using agonist and antagonist substances to presumptive neurotransmitters using in vitro and in vivo preparations.
- o Develop monoclonal and polyclonal antibodies to newly-identified neurotransmitters to determine their cellular localization by means of immunocytochemistry.
- o Study changes in gene expression in vestibular neurons during development, aging and plastic, adaptive alteration of vestibular function.

Understanding Pathophysiology

- o Acquire human tissues such as temporal bone and central nervous system structures from normal subjects and patients with documented vestibular symptoms and study the tissues by means of light microscopy, transmission electron microscopy, scanning electron microscopy and cytochemical methods.
- o Develop and study animal models to provide insight into diseases that currently are incompletely explained, such as vessel loop compression of the vestibular nerve and metabolic effects of endocrine disorders.
- o Study the effects of compromising the blood supply to vestibular sense organs. This study should be preceded by a description of the vascular supply and flow to the vestibular labyrinth.
- o Study the effects of specific vestibular lesions in animal models.
- o Encourage young histopathologists and provide training in peripheral and central vestibular system pathology.

Needs in Diagnosis

The overall objective of vestibular testing is to utilize an efficient battery of tests to determine the site and severity of a lesion. This battery should be a quick, efficient set of tests that minimize the time and cost to the patient and optimize the diagnostic value to the physician. Research goals in this area include the need to:

- o Develop standards, by age group, for the more commonly used diagnostic techniques including caloric, rotational and posturographic tests. No acceptable or standardized methods have been established for these tests and variations in the methods used in different centers make it difficult to compare results.
- o Use animal models and human subjects to develop new tests to evaluate all the recognized receptor organs in the vestibular apparatus. In particular, there is a need for simple and reliable tests of otolithic organ functions. There is a similar need for tests of vertical semicircular canal function.
- o Develop psychophysical methods for evaluation of vestibular function in health and disease.
- o Develop tests for alteration in neck and vestibular function and neck and vestibular interactions.
- o Use animal models to study the pathophysiology of disease

processes, such as Meniere's disease, vestibular neuronitis, positional vertigo, benign paroxysmal positional vertigo, unilateral and bilateral hypofunction and ototoxicity.

- o Conduct serial studies of changes in vestibular test results in patients with well-defined pathologic processes including evaluation of the process of compensation and resulting changes in reflexes.
- o Investigate the effect of pharmacologic therapy on the rate of recovery produced by compensatory processes.
- o Develop diagnostic procedures to evaluate vestibular developmental processes by the study of patients with developmental disorders and animal models of genetic disorders with abnormal receptor function, e.g., use of animal mutants without otoliths.
- o Investigate the pharmacologic, molecular and genetic basis of inner-ear disorders.
- o Develop biochemical and histological methods to detect immunologic, autoimmune and neoplastic abnormalities in the inner ear and eighth nerve.
- o Compare results of observations made in patients with central vestibular disorders to those made

in patients with disorders of visual and motor function such as congenital nystagmus, other abnormalities of ocular stability and ataxic syndromes secondary to cerebellar degeneration using brain stem evoked response, visual evoked response and neural conduction velocity tests.

- o Use postmortem material to confirm presumed diagnosis on longitudinally-well-studied patients; evaluate test results for the identification of a known cause of disease; study patients with unknown causes; and investigate immunologic and molecular changes in the inner ear and central nervous system of patients with vestibular disorders.

Needs in Treatment

Medical Therapy

The goals of current medical therapy are to relieve symptoms and ameliorate aberrant signals from an abnormal peripheral vestibular organ. Presently, all medicines which suppress vertigo and the associated autonomic symptoms have important side effects. They cause drowsiness, dry mouth and eyes and may interfere with normal central nervous system function. Therapeutic agents with greater specificity need to be developed. These developments should result from basic science experiments, for example

employing monoclonal antibodies to locate specific receptor sites within vestibular pathways. Work should be continued on the following topics, and specific goals are to:

- o Investigate neurotropic agents such as nerve growth factor and thyroid and adrenocortical stimulating hormones, showing some promise but not yet having been sufficiently evaluated to be recommended for clinical use.
- o Determine the possible deleterious effect of vestibulo-suppressive medications on rehabilitation.
- o Compare prospectively the effects of ablation with the effects of aminoglycosides.

Surgical Therapy

Work should be continued on the following topics, and specific goals are to:

- o Evaluate, in carefully controlled, clinical studies, the effectiveness of controversial procedures, such as vessel loop relocation, perilymph fistula "repair" and canal "plugging" for benign paroxysmal positional vertigo. Animal models with documented histopathologic correlates of these lesions should be employed as well.
- o Examine the anatomic and physiologic aspects of the central

compensatory events that follow labyrinthectomy and vestibular neurectomy in animal models. Short- and long-term assessment of these changes may help to determine the optimal therapeutic procedure.

Rehabilitation

Work should be continued on the following topics, and specific goals are to:

- o Determine whether exercise enhances vestibular rehabilitation and/or improves the level or rate of recovery in patients with vestibular disorders using prospectively-controlled studies which quantify changes in vestibular function. The exercise approaches used should be founded on knowledge of the normal anatomy and physiology of the vestibular system and may be specific for different disorders.
- o Develop outcome measures for assessing treatment efficacy, including postural responses.
- o Investigate the effect of co-morbidity on recovery following vestibular lesions, especially the effect of disorders that affect the visual and somatosensory systems.
- o Determine the adaptive capability of the vestibular system in young and old subjects and patients with various vestibular disorders.

Research Opportunities

Major Basic Scientific Opportunities

There are five broad, high-priority areas for basic vestibular research: signal transduction by vestibular end-organs; reflex control of posture and gaze; sensory integration in spatial orientation, perception and motion sickness; adaptive changes in vestibular function; and development and aging of the vestibular system. Within each area opportunities exist for research at the levels of: behavior, kinesiology and biophysics; anatomy and physiology; cellular properties, biophysics and metabolism; pharmacology, molecular biology and genetics; and mathematical and computer modeling. These opportunities can be summarized as follows:

Signal Transduction by Vestibular End-Organs

Important goals are to understand how vestibular end-organs convert head movements into neural signals and to determine the way in which this conversion is modulated by efferent projections from the brain to the end-organs. Research is needed to:

- o Describe the motions of the cupulae and otolithic membranes in response to head movements and examine the possible role of hair cell motility in modifying these motions.
- o Determine the distribution and organization of otolithic organ and semicircular canal inputs to the vestibular nuclei and cerebellum and elucidate the function of the vestibular efferent system in behaving animals through anatomic and physiologic research.
- o Characterize the biophysical properties of hair cells and afferent fiber terminals. This research should include studies of the hair cell transduction channel, the basolateral currents within hair cells and the efferent actions on both hair cells and afferent nerve terminals.
- o Conduct molecular biological studies which isolate and characterize the molecular components of the transduction channel and associated regulatory proteins.
- o Identify neurotransmitters which mediate peripheral vestibular function.
- o Investigate metabolic requirements of the sensory organs and neurons in the inner ear and the

mechanisms of production of inner-ear fluids and transduction elements (cristae and maculae).

- o Develop dynamic models that predict and explain patterns of afferent fiber activation on the basis of anatomical and biophysical properties of end-organs.

Reflex Control of Posture and Gaze

The goal is to understand the structure and function of neural circuits that transform vestibular sensory input into the motor output required for control of posture, locomotion, fine motor activities and gaze. Research is needed to:

- o Characterize the behavioral strategies and neural mechanisms used to determine posture and gaze during active and passive motion and during combinations of linear and angular acceleration in three dimensions. Studies are also needed to define the effect of exposure to altered gravito-inertial fields on vestibular reflexes and their potential clinical consequences.
- o Characterize the anatomic and physiologic properties of neuronal pathways that constitute the vestibular system in animal models and normal humans. It is especially important to identify pathways involved in vestibular reflex control of neck, axial and limb muscles and

to determine the signals they carry. Emphasis should also be given to understanding the neural substrates of otolithic organ reflexes and postural stabilization.

- o Correlate structure and cytochemistry of neurons in different vestibular circuits with their response characteristics.
- o Characterize the biophysical, molecular and pharmacological properties of specific groups of relay neurons in postural and gaze reflex pathways.
- o Develop multidimensional models that explore the neural basis of dynamic and spatial transformations in vestibular reflexes.

Sensory Integration in Spatial Orientation, Perception and Motion Sickness

Understanding of the vestibular system requires knowledge of how vestibular signals interact with information from other senses to generate perceptions, movements and motion sickness. Research is needed to:

- o Describe sensorimotor and perceptual reactions to complex combinations of linear and angular accelerations and determine how vestibular, visual and proprioceptive inputs interact to

generate perceptions of space and body motion.

- o Conduct anatomic and physiologic research to characterize the neural mechanisms that combine otolithic organ, semicircular canal, visual and somatosensory information to generate perceptual and postural responses and that generate motion sickness.
- o Characterize the neural and humoral mechanisms associated with motion sickness and vertigo at the cellular and molecular levels.
- o Develop models that incorporate and test the understanding of the neural basis of postural and gaze control during active and passive motion.

Adaptive Changes in Vestibular Function

Studies of vestibular adaptation can both reveal basic principles of motor learning and lead to strategies for enhancing recovery from vestibular lesions. Research is needed to:

- o Determine at the behavioral level, both the extent to which the adaptive system can compensate for vestibular dysfunction and the sensory cues that are important in producing this compensation. Adaptive changes in both rapid and slow components of the VOR and in postural reflexes should be studied.

- o Conduct anatomic and physiologic research to determine where in the brain stem and cerebellum the neural changes responsible for adaptive alteration of vestibular reflexes occur and how these changes lead to the observed alterations in behavior.
- o Characterize the biophysical changes that occur at various neural sites during the adaptive process.
- o Analyze the pharmacologic and molecular bases of adaptive changes and the way in which they depend on expression of molecular mechanisms such as proto-oncogenes, second messengers or humoral factors.
- o Develop models that account for adaptive changes at both the biophysical and neural circuit levels.

Development and Aging of the Vestibular System

Knowledge in this area will both help us deal with developmental disorders and age-related declines in vestibular function and also contribute to basic understanding of vestibular mechanisms. Research is needed to:

- o Characterize at the behavioral level the progression of perceptual and

reflex function during development and aging.

- o Conduct anatomic and physiologic research to determine the mechanism(s) underlying development of central vestibular pathways, their relation to development of peripheral end-organs and the ontogenetic sequences of connectivity between vestibular neurons and motor and higher sensory centers in the brain.
- o Study how membrane and synaptic properties of hair cells and vestibular sensory and motor neurons change during development and aging and examine the roles of neural activity and neurotransmitters in producing those changes at the cellular level.
- o Determine the neurotransmitters present in vestibular brain stem nuclei over the course of development, maturation and aging.

Major Clinical Scientific Opportunities

There are six broad high priority areas for clinical vestibular research: prevalence and environmental factors; anatomic, physiologic and molecular bases; diagnostic methods and testing procedures; adaptive mechanisms; and medical and surgical therapy. These

opportunities can be summarized as follows:

Prevalence and Environmental Factors

It is important to develop a better understanding of disease prevalence and possible associated environmental factors. Research is needed to:

- o Assess the distribution of balance disorders among different sectors of the population (including age, gender, genetic background and geography).
- o Identify environmental and occupational hazards that adversely affect balance.
- o Carry out a demographic study to evaluate the deleterious effects of prescription and over-the-counter medications which may produce central or peripheral vestibular alterations.

Anatomic, Physiologic and Molecular Bases

A better understanding of anatomic, physiologic and molecular bases of normal and abnormal balance processes is needed.

- o Determine modifications of neuronal vestibular pathways in animal and human pathologic specimens with known vestibular dysfunction.

- o Analyze the molecular and pharmacologic properties of specific groups of relay neurons in postural and gaze reflex pathways. The development of these properties from embryo to adult vestibular systems should be traced and changes that accompany aging examined.
- o Determine the coexistence of multiple neurotransmitters within single vestibular synapses.
- o Determine the excitatory and inhibitory neurotransmitters in the vestibular portion of the brain stem nuclei during development, maturation and aging.
- o Develop animal models to study the pathophysiology of vestibular diseases.
- o Examine structural changes in vestibular pathways in animal and human pathologic specimens with known vestibular dysfunction.
- o Investigate the molecular basis of acquired and congenital inner-ear disorders in humans and animal models.
- o Study and correlate peripheral and central pathologic abnormalities with biochemical and molecular changes in human subjects with well-studied disease.

Diagnostic Methods and Testing Procedures

Improved tests of balance function, a more standardized method of testing and research on pathology are needed .

- o Develop new tests to evaluate otolithic organ and vertical semicircular canal function in humans, including freely-moving subjects.
- o Develop tests to define the contributions of neck receptors to normal gaze and balance function and to vertigo and imbalance in pathological conditions.
- o Develop new tests for vestibular function involving non-invasive recording of neural activity and/or reflex responses elicited by specific mechanical or electrical stimuli that activate vestibular afferents.
- o Develop new and improved methods for the evaluation of posture under static and dynamic conditions.
- o Develop standards for the more commonly-used diagnostic tests in the vestibular test battery.
- o Develop improved psychophysical methods for evaluation of vestibular function in health and disease.

- o Develop animal models to study the reliability and validity of new vestibular tests.
- o Develop methods to detect immunologic and autoimmune inner-ear disorders and neoplasms for the diagnosis of vestibular system disorders.
- o Validate currently accepted diagnoses or new diagnoses based on clinical pathologic correlation.
- o Conduct serial (longitudinal) studies of the changes in vestibular tests in patients with well-defined pathologic processes.
- o Determine if there is a critical period for the initiation of vestibular exercises to facilitate recovery following the sudden onset of vestibular loss or dysfunction in humans.
- o Determine the optimal characteristics of vestibular stimulation necessary to facilitate recovery in different vestibular disorders.

Medical and Surgical Therapy

Medical and surgical therapy is frequently based on insufficient determination of efficacy. Standardized reporting of results is critical, and the efficacy of treatment should be determined.

Adaptive Mechanisms

Adaptive mechanisms play an important role in normal balance function, compensation for disease and rehabilitation.

- o Analyze the molecular and pharmacologic bases of adaptive changes in the vestibular system.
- o Investigate the use of pharmacologic agents to modulate changes in adaptive compensation.
- o Extend studies of adaptive behavior to consider the conditions of unilateral and bilateral labyrinthectomy in an attempt to understand the response to vestibular damage.
- o Perform multi-center, prospective, controlled clinical trials which:
 - o evaluate medical therapy for vestibular symptoms.
 - o evaluate osmotic or renal loop diuretics for the treatment of episodic vertigo.
 - o compare medical to surgical ablative therapy in Meniere's disease.
- o Study the pathologic effect of vessel loop compression on the vestibular nerve and the effectiveness of vessel loop relocation.
- o Study the anatomic, physiologic and pharmacologic central nervous

BALANCE AND THE VESTIBULAR SYSTEM

- system correlates of neurectomy (partial and total) compared to labyrinthectomy in appropriate animal models.
- o Determine the effect of surgical manipulation of the cochlear nerve and the facial nerve intracranially and in the internal auditory canal.
- o Determine the effect of the peripheral and central molecular, biochemical and structural changes following various ablation procedures, e.g. labyrinthectomy, and partial and total vestibular neurectomy.

**LANGUAGE AND
LANGUAGE IMPAIRMENTS**

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LANGUAGE AND LANGUAGE IMPAIRMENTS

Overview

Language is the expression of human communication through which knowledge, belief and behavior can be experienced, explained and shared. The ability to manipulate language to satisfy needs and desires and to express thoughts, observations and values is an important human pursuit that directly influences the quality of life for any individual.

The broad goals of research on language are to understand the nature of normal language function, including the underlying bases and mechanisms involved. One goal of this work is to build the foundation necessary to develop and evaluate intervention and rehabilitation strategies to improve and enhance the communication process for individuals with language disorders. The understanding of normal language (whether spoken, signed or written) provides a basis for comparison in investigations of language disorders. It is critical to understand how language is produced and understood, what its biological and neural substrates and organizing principles are, how it is

learned by children and how it is processed.

For an adequate understanding of language functioning in children and adults, research efforts must include all of the diverse groups that make up contemporary United States society. These populations include racial and ethnic minority groups and groups categorized in terms of gender, age, geographic region and social and economic status.

In the United States, there may be as many as one half million persons who were born deaf or who lost their hearing before they acquired spoken language. In the world at large the number may approach 10 million. A large proportion of these individuals use a form of signed language as their primary mode of communication. Some use spoken English exclusively. Many use both. Research and services related to deafness must be concerned with the impairment of auditory language and with the status of and access to signed or spoken languages which are perceived visually.

There exists another group of at least one million people whose hearing impairment is less severe but was

acquired during childhood. Most of these individuals use spoken language as their primary mode of communication, although some also use a signed language. Despite the substantial benefit of auditory input to such people, their language acquisition is often characterized by difficulties not faced by normal hearing people.

Individuals with normal hearing, as well as those with a hearing impairment, may exhibit a disorder of language, that is, a deficit of language comprehension, production or use sufficient to impair interpersonal communication. In young children, these disorders frequently involve difficulty in the acquisition of the ambient spoken or signed language and may also lead to impairment in reading and writing. In adults and older children, impaired persons include aphasic individuals who have lost their previous levels of language competence as a result of brain injury.

Language impairments impede economic self-sufficiency, academic performance and employment opportunities. It is estimated that between six and eight million individuals in the United States have some form of language impairment. In addition to loss of livelihood, these disorders impose social isolation and personal suffering on the affected individuals and place an enormous emotional and economic burden on their families and on society as a whole. These disorders have a life-long impact on the ability of those affected to

make their way or even to survive in our technologically-advanced society.

Disorders of language affect children and adults differently. For the child who does not use language normally from birth or who acquires the impairment in childhood, the disorder occurs in the context of a language system that is not fully developed or acquired. In contrast, damage to the language apparatus in adults disrupts a system that is less malleable in the face of neural damage. Adults with aphasia commonly have highly-selective deficits and more highly-developed compensatory mechanisms. Neurologic, physiologic and metabolic differences between children and adults provide particular problems and challenges to the study of language disorders in these populations. As a result, while the broad goals of research on language disorders are similar for children and adults, the research agendas for these two groups are considered separately.

Recent Accomplishments

Multicultural Issues

The majority of children from all cultures acquire language with little difficulty, including those who are reared in multicultural environments. However, the assessment of disorders within

multicultural groups can be complicated by a variety of factors. For example, individuals who are members of multicultural populations such as African-, Asian- and Hispanic-Americans may be incorrectly identified as language impaired due to the use of culturally-inappropriate language assessment instruments. On the other hand, such groups are more likely to live in conditions of poverty, which give rise to various health and social conditions that are linked to increased risk of communication disorders.

There are likely to be pertinent differences in prevalence, causes and manifestations of language disorders among diverse cultural populations. For example, sickle cell anemia, a condition associated with an increased prevalence of sensorineural hearing loss and neurological impairment, is estimated to occur in one in every 500 African-American children. To the extent that these complications directly influence language learning, such children are at considerable risk. Current data also show a very high prevalence of chronic middle ear disease among Pacific Rim, Alaskan Native and Native American populations. The prevalence of hypertension and diabetes, two of the primary risk factors for stroke, may be higher among low socioeconomic groups. Thus, there is likely to be an increased occurrence of aphasia in these groups.

Language disorders have been found to occur in children with elevated blood lead levels. The highest blood lead

levels have been found in children living in low-income households within the inner cities of large urban areas.

Language Among Deaf Children and Adults

Language acquisition takes place naturally for most children; those who have normal hearing and those who are deaf with signing deaf parents. Many deaf children use a natural sign language, American Sign Language (ASL), which shares an underlying organization with spoken language. Recent electrophysiological findings show that, in spite of the very different input/output systems employed, the same areas in the left hemisphere of the brain are involved in language tasks in native signers and speakers of English. However, despite the ease with which they acquire ASL, many deaf children of deaf parents have inordinate difficulty learning to read and write.

Studies of the acquisition of ASL suggest that signers acquiring the language at a later age demonstrate grammatical deviations from more standard ASL. These deviations persist even four or five decades after acquisition has taken place. This finding suggests the existence of critical periods for acquiring signed language that parallel those documented for spoken language.

Studies of deaf children of hearing or oral deaf parents indicate that, with

intensive oral training, their acquisition of spoken language and reading tends to be superior to that of deaf children with equivalent levels of hearing impairment who do not have such training. However, deaf children of hearing or oral deaf parents often display substantial delays learning to read and write.

The dominant pedagogical methodology employed currently in the United States in teaching language to deaf children involves the exposure to artificial signing systems, which are intended to represent and model English and to promote the natural acquisition of English grammar. Recent research suggests that deaf children of hearing parents, when exposed only to artificial signing systems, develop some idiosyncratic grammatical patterns that may not reflect the structure of English or of ASL.

Another methodology currently employed emphasizes the development of spoken language exclusively. Some deaf children exposed to this method throughout their educational program achieve English language competence without the knowledge of sign language.

Cued Speech, a system of communication which uses simple hand cues in conjunction with the natural mouth movements of speech, emphasizes the natural development of spoken language. Many deaf people exposed to this system throughout their educational history achieve English language competence.

Just as speech is one of the basic building blocks of language for hearing people, signing is one of the basic building blocks of language for some deaf people. Thus, the study of sign language perception is as critical to an understanding of the language of many deaf people as the study of speech perception is to understanding the language of hearing people and deaf people who use auditory and/or visual means for perceiving spoken language. Such studies will provide insight into the nature of language processing in deaf people and will lay the foundation for understanding disorders of spoken language that may occur in certain deaf persons. Comparisons of the processes of spoken language perception and signed language perception in normal and language-disordered hearing and deaf individuals provide a unique means of determining those aspects of language that are independent of the mode (signed or spoken) of communication. Such findings may be used to develop appropriate rehabilitation strategies, depending upon the nature of the language deficit.

Studies of the acquisition of sign suggest that infants are very good at relating information in one sensory modality with information in another. Deaf infants learning a signed language rely on movement of the hands and arms, as well as processing by the eye. New technology for three-dimensional motion analysis has been developed for the study of signed language perception, and investigations are now under way that

will allow one to separate constraints imposed by the transmission modality from more centrally determined factors in the perception of the basic building blocks of language.

Central to the understanding of disorders of spoken and signed language perception in the hearing impaired is an understanding of normal processes. Research advances in the past 10 years have focused primarily on spoken language perception in normal persons and while many advances have been made, scientists are only beginning to understand fully the nature of the spoken and signed language perception process.

For most people, spoken or signed language, or a combination of the two, are the ways to express thoughts and ideas and to communicate with each other. How spoken or signed language perception interacts with syntactic and semantic knowledge in language comprehension is still not understood. The increased availability of precise instrumentation techniques and use of more sophisticated research methods promise to provide a richer understanding of the nature of spoken and signed language perception. Largely through the efforts and cooperation of many different kinds of scientists (speech, language and hearing scientists, engineers, linguists and psychologists) research on this topic is beginning to contribute a multi- and inter-disciplinary perspective to this complex but critical problem.

Language and Language Disorders in Children

The study of language acquisition in normally-developing hearing children has provided an important foundation for research with language-disordered children. Findings on both the course and underlying bases of language development have been reported. It is clear from this work that infants have available at birth, or quickly develop, many of the perceptual, cross-modal (auditory-visual) and conceptual abilities necessary to learn language. Differences among individual children's language learning patterns have been identified, as well as differences in learning according to the type of language being acquired (e.g., morphologically-rich like Italian vs. word-order-dominant like English). Although much research remains to be done, the existing knowledge has greatly facilitated the study of language impairments.

Language disorders among hearing children can be discussed in terms of whether the language difficulties exist in isolation or in association with other problems and whether the factors interfering with language were present from birth or appeared subsequently.

Many children with isolated language problems that appear to be present from birth are given the clinical label of specifically language impaired (SLI). These children show normal hearing, age-appropriate scores on

standardized tests of nonverbal intelligence and no overt evidence of neurological damage. Although U.S. prevalence data need to be more firmly established, it is estimated that approximately five percent of preschool children fall into this clinical category.

Although SLI children do not show signs of frank neurological impairment, neuropsychological studies have revealed that these children perform poorly on perceptual and memory tasks, especially those involving the processing of rapid acoustic changes. These findings cast doubt on the presumed isolation of the language difficulties.

Investigations of the language skills of SLI children have focused on syntax, morphology, phonology and semantic relations. These studies have revealed significant limitations in each of these areas. Although each area is acquired in a manner approximating normal development (albeit more slowly), the language profiles across areas often do not match those of younger, normally-developing children in that certain areas (for example, morphology) may show an especially serious deficit. Because studies have focused exclusively on SLI children acquiring English, it is not known whether the observed profiles reflect general difficulties with particular grammatical functions or are influenced by the manner in which these properties are marked in English. Several retrospective, follow-up studies of SLI children have suggested that residual problems with language and language-

related learning problems may be seen through adolescence and into adulthood.

Research in recent years has begun to address the problem of inadequate subject description of SLI children. This work has provided a protocol for selecting prototypic groups of SLI children. Projects devoted to distinguishing among subgroups of SLI children are now under way.

Efforts aimed at the early identification of SLI children are also in progress. It appears that children's expressive vocabulary size, vocabulary comprehension and use of symbolic gestures assist in determining whether late-talking children are at risk for language impairment.

New technologies such as magnetic resonance imaging (MRI) and event related potentials are beginning to be applied to SLI children. Research to date suggests that SLI children show an atypical left-right hemispheric configuration. Genetic studies of SLI children have also been undertaken. The preliminary findings from these studies suggest that SLI children are more likely than normally-developing children to have other members of the family with present or resolved language difficulties.

Research on the phonologic, morphologic and syntactic features of language that are difficult for SLI children, as well as on the perceptual and motor abilities of these children, has continued. In addition, the lexical

(vocabulary) and pragmatic (communicative-conversational) abilities of SLI children have received investigative attention.

Prospective longitudinal research on SLI children has begun. The results of these studies indicate long-term deficits in these children. Evidence is accumulating that young SLI children are clearly at risk for later deficits in reading and that SLI and reading-disabled children represent overlapping populations.

For many children with language disorders, limitations in other areas are also evident. Some of these multiple disabilities result from maternal substance abuse in pregnancy, fetal and infant malnutrition, lead poisoning, congenital AIDS and prematurity in the children of adolescent mothers without prenatal care.

In terms of prevalence, mental retardation is the most common disorder associated with inadequate language development. Recent research shows that children with mental retardation can have a variety of language disorders and that mental retardation often offers an inadequate explanation for communication problems in these children. A common genetic form of mental retardation, Down syndrome, has been shown to be associated with greater impairment of expressive than receptive language. Fragile-X, probably the most common single cause of genetic mental retardation in males, affects language

meaning and use more than it does the acquisition of phonology and syntax.

Autistic children constitute another group in whom language is but one area of deficiency. Inadequate communication skills are hallmarks and the most common presenting symptoms of autism, but nonlinguistic deficits make separate contributions to its symptomatology. Nonetheless, it is now clear that, while autism is associated with mental retardation in some children, mental retardation is not a defining feature of the disorder.

Other conditions that can complicate the normal acquisition of language relate to the availability of an adequate listening environment early in life. This need is illustrated by three findings. First, there are some data to suggest that chronic otitis media in the developing infant, when accompanied by a mild and intermittent hearing loss, may be predictive of later language impairment. Second, there is evidence that early amplification and auditory training can significantly affect the deaf child's acquisition of spoken language. Third, there is evidence that normal infants and children need a more favorable signal relative to the background noise to perform at the same level as the adult in spoken language perception tasks.

Some language disorders in children are acquired through infections, tumors, stroke or trauma. Strokes in children are estimated to have an annual

incidence of 2.52 per 100,000 children. Other causes, such as head trauma are estimated to be as high as 200 per 100,000 per year.

Advances have been made in specifying the relationship among acquired language deficits and the focality or diffuseness of central nervous system (CNS) involvement, lesion laterality and age at lesion onset. Children with focal, unilateral lesions, such as those sustained following vascular episodes, generally have been found to have better long-term language development and recovery than children with lesions involving more diffuse brain structures, such as CNS tumors treated by whole head radiation and chemotherapy, severe closed head injury or epileptic aphasia. Both fluent and nonfluent aphasia may occur in children with acquired brain lesions, and a variety of syntactic and lexical comprehension and production deficits have been described following left hemisphere lesions. Delays in lexical development and in the development of syntactic structures have been documented following early lesions of either the left or right hemisphere. Attempts to relate language sequelae to lesion location within a hemisphere have been equivocal. Prognosis related to age at lesion onset is controversial and confounded by factors such as the diffuse nature of the lesion, concomitant seizure disorders and questionable premorbid status. Studies using electrophysiologic and neuroimaging techniques are beginning to address the nature of hemispheric

reorganization following acquired language loss in children.

Language and Language Disorders in Adults

Current best estimates place at nearly one million the number of adults in the United States with acquired disorders of language due to stroke or traumatic brain injury. Additionally, a large proportion of the estimated two million citizens with progressive dementing disease have significant language impairment. Their disabilities range from partial impairment that affects primarily one or two input or output channels to near total and permanent loss of comprehension and production of speech. While many are rendered totally dependent, there is a wide range of possibilities for rehabilitation of communicative power and, in some cases, the return of economic self-sufficiency.

Impairments in the comprehension, production or use of language by adults are encountered in a variety of clinical settings. Acquired language disorders are frequently observed, for example, in patients with stroke, head injury, dementia, brain tumors and CNS infections (including AIDS). Language disorders are also found in adults who have failed to develop normal language because of childhood autism, hearing impairment or other congenital or acquired disorders of brain development. Although deficits of spoken language frequently affect all language modalities,

that is, reading, writing and the signed languages of deaf people, dissociations in performance as a function of language modality do occur. Thus, for example, patients with acquired disorders of reading or writing may be essentially normal in spoken language. Lastly, disorders of language, or communication more generally, may be encountered in patients with dysfunction of the non-dominant hemisphere. Specific impairments in the interpretation of communicative intent, as well as in the ability to appreciate several alternate meanings of a word, have been demonstrated after stroke involving the right hemisphere.

Research on the ways in which adult language can break down following brain injury necessarily builds upon an understanding of how language comprehension and production are accomplished by normal people. The development of experimental techniques for analyzing language functions and for investigating the neuroanatomic representation of those functions in the brain constitutes an important component of current efforts in this area.

The understanding of the anatomic and physiologic bases of normal and disordered adult language has, in recent times, been facilitated by the application of a variety of experimental techniques. Contemporary computed tomography (CT) and magnetic resonance imaging (MRI) scans offer precise information about lesion size and location heretofore available only at autopsy. These data

have, for example, contributed to a better understanding of the anatomic basis of speech initiation and production. Imaging of brain metabolism and blood flow using positron emission tomography (PET), as well as single photon emission computed tomography (SPECT), has contributed substantially to the understanding of the functional anatomy of the language system in normal subjects as well as in patients with brain dysfunction. These studies of dynamic brain activity have also led to a greater appreciation of the important interactions between language and other cognitive operations. Electro-cortical stimulation techniques have also contributed to the understanding of the functional anatomy of the language system. The analysis of the consequences of transient electrically-induced cerebral "lesions" has facilitated the identification of discrete language mechanisms and has engendered better understanding of the individual variability in the cortical representation of language functions. Finally, additional electrophysiologic techniques, such as event-related potentials (ERP) as well as advances in the mathematical modeling and signal processing of electroencephalographic data, have assisted in the understanding of the temporal course and anatomic representation of language processes.

The theories and methods of modern cognitive science have brought about important refinements in understanding the cognitive processes that normally underlie language functions. For example, chronometric

investigations of the course of auditory language comprehension have highlighted the complexity of the processes required to integrate aspects of word meanings with elements of sentence structure. These new insights have motivated the development of tests that allow the attribution of specific aphasic symptoms to failure within identifiable components of the language system. These advances have, in turn, provided the groundwork for the development of new approaches to diagnosis and rehabilitation of aphasia and acquired dyslexia.

Several recent findings await further study and fuller exploration. For example, common mechanisms underlying spoken language and the signed language of deaf people have been dramatized by the identification of striking parallels in the effects of brain injury on these two modes of communication. Artificial intelligence computer models of language have offered insights into the processes mediating language; additionally, disruptions or "lesions" of these models permit the simulation of symptoms and promise to provide a means for the testing of hypotheses concerning the basis of particular language deficits. Cross-language comparisons of sentence production and sentence comprehension disorders have begun to distinguish between symptoms that are universal in their form of presentation and those that are specific to the structure of particular languages.

Recent advances in the remediation of language disorders include the demonstration that some profoundly-aphasic patients can learn a computerized system for exchanging information by manipulating visual symbols. In fact, computer-assisted assessment and instruction are active areas of current research interest and promise to allow the testing of previously-untapped cognitive capacities in severely-impaired patients. Explorations in the pharmacologic treatment for aphasic symptoms have shown promise for the relief of selective disorders, such as impairments of speech initiation, through the use of the dopamine agonist, bromocriptine.

Several more traditional treatment programs have been shown to be efficacious with specific types of aphasic patients. These include language oriented and language stimulation treatment techniques as well as interventions designed to bring about well-defined outcomes such as the elimination of the perseverative intrusions of earlier utterances. Additional intervention strategies currently being tested include treatment protocols targeted directly at theoretically-defined language components that are found to be impaired. A limited number of such studies conducted to date have demonstrated measurable improvement among individual aphasic patients many years following onset of their aphasia. These studies represent a direct application of results gathered previously

in cognitive and linguistic studies of aphasia.

Recent accomplishments in the area of adult language and its disorders have built upon advances in neuroanatomic and neurophysiologic diagnostic procedures, as well as on developments in linguistic theory and cognitive science. Assessment and rehabilitation of these deficits continue to rely on the clinical expertise of speech-language pathologists with new participation from the fields of neuropsychology, pharmacology and computer science. This diversity reflects the complex and multifaceted nature of adult language disorders and provides a broad base upon which to develop a research agenda for the future.

Program Goals

A coherent program that addresses the topic of language and its various impairments must build upon previous accomplishments. At the same time, such efforts should fully exploit emerging technological advances and should accommodate widespread social changes that are likely to affect the acquisition of language and its use.

Several general themes are evident in the research priorities that can be developed to address specific impairments of language. One compelling requirement for improving

our understanding of and responses to language impairments is comprehensive data relevant to the incidence and prevalence of these impairments in the population. Systematic study of the epidemiology of these disorders, together with detailed information about the medical, social and cultural conditions with which they occur, would provide a foundation for approaching the important program goal of prevention of language impairments. A second basic requirement that is crucial to understanding any language impairment is detailed information about how language (in all of its auditory and visual manifestations) is acquired and used by normal individuals. The development of theoretically-based, methodologically-rigorous and culturally-unbiased test instruments, as well as the gathering of comprehensive normative data using such instruments, is central to this effort. In light of the growing role that diverse racial, ethnic and social groups play in society, it is essential that these populations be properly represented in the subject pools for studies of normal language processes.

Many language impairments occur because of brain injury or because the nervous system does not develop normally. A better understanding of the mechanisms by which the human brain acquires and processes language would greatly aid in the prevention and rehabilitation of many types of language impairments. Consequently, the exploration of newly-emerging

technologies for relating language functions to brain regions is a priority.

In addition to these general program goals, areas of specific concern can be enumerated.

- o Research on the language of deaf people requires particular attention to the specific needs of deaf children acquiring language in different learning environments. The effects on spoken and signed language development of early exposure to spoken English, signed English and ASL needs to be determined. The effects of differing levels of auditory information on spoken language development need to be rigorously assessed, particularly with regard to improvements provided by new assistive devices. Special attention should be directed to the factors that relate to the acquisition of literacy by deaf people, with the goal of developing methods for enhancing the acquisition of written language by improving English language instruction and by exploiting the child's existing language capacity.
- o Basic research on the structural characteristics of signed languages is needed to enhance comparison with information about spoken languages. In addition, continued investigation of differences and similarities in the way the brain

processes spoken and signed languages is needed.

- o Research on how signed languages are perceived and learned would contribute important information on the development of methods for teaching signed languages to deaf and hearing individuals.
- o Studies of children with language disorders should be devoted to several types of issues. Information is still needed on the linguistic, motor, neurogenic and cognitive bases of language disorders in children.
- o Valid and reliable language assessment tools are still needed. There is an especially critical lack of assessment instruments for children from multicultural populations.
- o The long-term consequences of language impairment warrant further study. Although it is well established that language disordered children are at risk for later academic, social and vocational difficulties, the nature and degree of these relationships must be determined with greater precision.
- o There is a pressing need for research on language intervention and treatment efficacy with language-disordered children. Although studies to date indicate

that treatment is effective, many specific questions (for example, the criteria used to match particular children with particular treatment procedures) are unanswered.

- o A fuller understanding is needed of how language in all of its many aspects is related to the anatomy and physiology of the brain. Relevant data may come from lesion studies seeking to map specific language capacities to brain regions and assessments of the functional anatomy of language in normal and brain-injured subjects using current imaging and electrophysiologic techniques.
- o The identification of the component processes underlying normal language remains a high priority. Investigations motivated by current theories of language processing of normal subjects employing spoken, written and signed languages, as well as detailed studies of patients with language deficits, will be relevant.
- o Improved understanding of the structure of normal language, as well as technological advances in computer applications, provide a basis for the development of innovative means for evaluating the language-impaired individual's symptoms and for addressing the deficits that are uncovered. Critical to this endeavor will be the development of experimental

designs to evaluate the utility of the assessment instruments and the effectiveness of new and existing treatments.

- o Finally, a rich source of data about language organization and potential breakdown is available from the comparison of the various manifestations of language impairment. Comparative studies can reveal mechanisms common to different causes and should provide a rational basis for diagnosis and treatment.

Research Opportunities

Multicultural Issues

Although United States society is increasingly multicultural, little research has focused on the implications of social and cultural differences for language and its impairments. Future investigations should:

- o Include normative studies for different languages or social dialects which focus on the acquisition and use of phonological and grammatical forms as well as content and pragmatics. Investigations of the simultaneous or consecutive acquisition of two or more linguistic systems are also

necessary. Such research can include investigations of "critical periods" for dual language learning and code switching.

- o Obtain basic epidemiologic information on the incidence and prevalence of and risk factors for speech, language and hearing disorders within African-American, Hispanic, Asian/Pacific Islander and Native American populations, as available data may not be generalizable across these groups.
- o Explore factors that might impede the efficacy of treatment, such as differences in cognitive or learning styles and cultural systems of belief.
- o Develop means of differentiating between language differences and disorders across cultural groups.
- o Investigate the incidence and prevalence and risk factors for adult neurogenic impairment among culturally-diverse and bilingual populations.

Language and Deaf People

The research needs for language and deaf people fall into three broad areas encompassing study of language development in children, literacy in deaf children and adults and basic research on sign language structure and function.

Language Development in Deaf Children

There is some evidence that, if other variables are held constant, deaf children of deaf parents may tend to outperform other deaf children on a variety of academic and psychosocial measures. A full explanation of this phenomenon is lacking and requires further study.

Studies are needed of normal patterns for acquisition of ASL and their relation to cognitive and psychosocial development among deaf children of deaf parents. In addition, to understand normal patterns of such development, it is necessary to pursue parallel studies of disordered courses of development of signed language in the same group.

Because deaf children of deaf parents constitute only a small percentage of the population of deaf children (between 5 and 10 percent of the total), it is imperative that a better understanding of typical patterns of language development of deaf children of hearing parents be pursued.

Within this area, new studies are needed to:

- o Identify and describe patterns of acquisition of ASL, particularly when access to a first natural language is delayed or incomplete.
- o Identify and describe patterns of language acquisition of children exposed primarily to systems or

styles of signing in which the intention is to model or support English speech.

- o Identify and describe patterns of language acquisition among children exposed primarily to oral language.
- o Identify and describe patterns of language acquisition of children primarily exposed to spoken English through Cued Speech.
- o Identify and describe patterns of related cognitive, psychological and academic deficits accompanying delayed language acquisition.
- o Study the nature and value of language models provided by hearing adults in contact with deaf children. This research should include studies of the nature of receptive and productive aspects of English-like signing in addition to aspects of auditory, oral language models.
- o Identify deaf children who have language disorders by examining and describing the differences between normal and language-impaired deaf children.
- o Study approaches and techniques for the training of hearing parents of deaf children to sign or to facilitate the development of their

children's spoken language, including the use of Cued Speech.

- o Identify and describe the processes of reorganization of the input language, particularly pidginization and creolization processes in language contact situations.
- o Identify and describe the invention of gestural language systems without formal sign input and the relation of this process to eventual signed or spoken language development.
- o Study approaches and techniques for evaluating the potential of new technology and intervention strategies (such as auditory prostheses, laser disk technology, computer-based training systems) for promoting spoken language acquisition by deaf children.
- o Identify critical periods for both the natural acquisition and training of spoken or signed language by deaf children.
- o Develop the means of identifying language disorders in deaf children from different cultures.

Studies of Literacy in Deaf Children and Adults

Because problems associated with the acquisition of reading and writing skills are the most pervasive academic

and vocational consequences of deafness, studies of the acquisition of English literacy must be included as a primary research objective. Research should be conducted to:

- o Study the nature of processing text by successful deaf readers in an attempt to identify the most effective strategies, both auditory and visual, for the teaching of literacy to deaf people.
- o Develop and test new strategies and techniques for teaching English as a second language to deaf children who make use of their capabilities for visual language processing.
- o Study the role of captioning, telecommunication devices, personal computers and other technological influences on the improvement of deaf children's literacy skills.
- o Study the relationships between speech processing and literacy in deaf children who use spoken language primarily.
- o Study the relationships among speech processing, sign processing and literacy in deaf children who use sign language.
- o Develop and evaluate appropriate and psychometrically-sound tests of psychosocial, intellectual and academic development and of first- and second-language acquisition

by deaf children and adults under a variety of language use conditions.

- o Examine the interaction of acquisition of signed languages with spoken and written language and begin to characterize the the resulting bilingualism. Studies of the order of acquisition of speech and signed language as they affect children's eventual acquisition of English language and literacy should also be conducted.

Basic Research on Sign Language Structure and Function

Studies of the organization of the brain and its relation to language function, especially signed language, are needed. Research on how signed language acquisition interfaces with other biological and physical systems is also important. Investigators should be encouraged to:

- o Continue research on brain mapping for sign language functions.
- o Develop improved techniques for imaging sign language motions in three-dimensional space.
- o Conduct further investigations into how deaf people perceive and process visually. Studies could include investigations of the relation between limb control and vision and of the functioning of deaf people with visual disorders.

- o Conduct studies of the nature of parallel processing of language, such as the simultaneous processing of visual and auditory information.
- o Expand studies of the brain and language, such as those on the specialization of the cerebral hemispheres for language and other cognitive processing.

Investigations of signed language structures are also important. Specifically, there should be attempts to:

- o Study signed language and other languages used by the deaf community from functional/psychological and structural/linguistic perspectives. Studies should focus on the nature of the "creolization" of signed language and the effect of interaction of modality and language structure, including contact signing.
- o Study signed languages other than ASL and varieties of signing within ASL, to identify the role of cultural and ethnic differences on signed-language use by deaf people.
- o Study other signed languages, such as simultaneous communication and signed English, relative to the acquisition of English language by deaf children.

The acquisition of the ability to employ a signed language depends on the development of a number of interrelated cognitive and linguistic abilities that contribute to the perception of sign. Interdisciplinary studies should be carried out to:

- o Relate infants' early acquisition of sign language phonology, assessed through tests of sign perception, to the acquisition of the higher levels of language, such as the acquisition of words (lexicon), word meanings (semantics) and grammar (syntax).
- o Investigate basic and higher level processes underlying vision, including the requirements of structured use of space and movement, the processing of complex dynamic arrays and the perception of motion and form.
- o Study the impairments to sign perception in deaf individuals who become aphasic or who sustain other cognitive impairments subsequent to brain damage.

Language and Its Disorders in Children

A number of questions pertaining to normal language development in hearing children are not yet resolved. Although the sequence of language attainments is generally known, at least for English, the bases of some of the acquisition patterns are not yet fully

understood. For example it is not yet clear the degree to which young children's early sentences involve abstract grammatical categories as opposed to semantically-based notions. The degree to which language learning mechanisms are modular rather than dependent upon more general cognitive operations must also be determined. Issues requiring further investigation can be found for each developmental period.

The research needs for language disorders in children fall into four broad areas encompassing study of the bases of language disorders in children, investigations of assessment, studies of academic, social and vocational impact and investigations of intervention.

Bases of Language Disorders in Children

Many questions concerning the very nature of language disorders remain unanswered. Certainly, discovery of the bases of language impairment is necessary before meaningful steps toward prevention can be taken. Similarly, identification of these bases and their accompanying symptoms is required for the development of an accurate typology for language disorders in children. Accordingly, it is essential to investigate a range of factors that may be implicated, including characteristics of the language being acquired and factors that influence the production of speech. The language characteristics themselves should be examined with an eye toward how they contribute to our

understanding of the child's difficulty. Research should be carried out to:

- o Examine the linguistic profiles of language-impaired children to determine if they vary according to the type of language being acquired (e.g., morphologically rich vs. sparse; flexible vs. rigid word order). The weaknesses observed in language-impaired children across these languages might reveal a common factor that may be the source of the disorder.
- o Examine the impact on language production of structural anomalies of the speech mechanism, such as craniofacial anomalies, long-standing tracheotomies or severe neuromotor impairment. The relative contribution of subtle motor speech problems to certain subtypes of language impairment is not known.

The neurogenic basis of developmental language disability is unknown in the vast majority of those with specific language impairment, as well as in those in whom these disorders are associated with other disabilities such as autism and mental retardation. The mechanisms by which neurogenic dysfunction occurs and the many genetic and acquired causes of developmental language dysfunction are still unresolved. A wide variety of intrinsic and/or extrinsic factors may be responsible for an initial induction of a

neurogenic abnormality. Research should be conducted to:

- o Study the normally-developing brain across the life span using a wide range of anatomic, physiologic and metabolic techniques. New advances in imaging the brain (anatomic, physiologic and metabolic) are particularly important to apply to studies investigating normal and abnormal development.
- o Conduct multidisciplinary studies that combine imaging techniques with fine-grained behavioral analysis of processes underlying language development and disorders.

Advances in molecular genetics, coupled with recent evidence of familial transmission in some cases of developmental language and learning disability, suggest that further genetic studies would be particularly profitable. Studies should combine state-of-the-art behavioral assessment with molecular procedures to:

- o Determine the reason for the much higher prevalence of developmental language disorders in boys than girls.

The precise role of nonlinguistic cognitive factors must also be examined in children with language disorders. Although many language-impaired children exhibit limitations in intelligence

as it is broadly defined, it is likely that particular aspects of cognition are more important to language learning than others. For example, many studies of SLI children have found deficiencies in both symbolic and nonsymbolic domains of knowledge even when scores on standard intelligence tests are within the normal range. Further work is needed to:

- o Isolate those cognitive areas not represented in IQ tests and determine whether or not these areas interact with language functioning.
- o Determine to what extent language impairment may be secondary to psychosocial disorders.
- o Determine to what extent psychosocial disorders may result from a primary language impairment. There are special groups of children and youths who are at risk for language disorders and language-based learning disorders. These special groups include victims of abuse, neglect or drug- or alcohol-related lifestyles and homelessness and general victims of poverty.
- o Determine to what extent failures in communication and education are related to psychosocial factors.

Assessment

The purposes of assessment include the identification of a language

impairment as well as the determination of the nature and severity of the impairment, the prognosis and the initial goals for intervention. Initiatives are needed to:

- o Develop psychometrically-sound and culturally-fair and sensitive procedures for the measurement of language comprehension and production to be used in identifying and classifying language impairments.
- o Develop alternative assessment strategies including criterion-referenced testing, observational techniques, interview procedures and other informal strategies.
- o Explore the utility of state-of-the-art computer technology in language assessment, including language sampling analysis.
- o Develop assessment strategies which have application, validity and reliability for identifying language disorders in children in a variety of assessment contexts (e.g., school, home, clinic).
- o Determine the most appropriate strategies for evaluating limited-English-proficient (LEP) children including the use of interpreters or informants.
- o Develop assessment tools for preschool populations, especially

for children under three years of age.

Academic, Social and Vocational Impact

The negative academic, social and vocational consequences of language disorders in children have been well documented; however, research is needed to:

- o Study the relationship between language disorders and subsequent or concomitant learning disabilities.
- o Study the relationship in children between language disorders and social adjustment problems.

Intervention

New intervention strategies for language-impaired children need to be developed that are capable of testing and refining current theoretical models. These approaches should also be tailored to meet the needs of individual children; that is, they should address documented, underlying perceptual and motor deficits as well as linguistic inadequacies. Furthermore, the strategies should be culturally relevant and salient to the individual child's experiences and future aspirations. Research must be conducted to:

- o Develop new intervention strategies based upon current

theoretical models and establish their efficacy.

- o Determine the efficacy of currently available intervention strategies through both single subject studies and studies of children who clearly represent different types of language impairment.
- o Evaluate alternative models of service delivery such as family- or school-based programs versus direct delivery by speech-language pathologists.
- o Investigate the use of computers and recent technologic advances (for example, in speech recognition and speech production) in intervention with language-impaired children and with deaf children learning oral language.

Language and Its Disorders in Adults

The research needs for adult language disorders fall into four broad areas encompassing research on language and the brain; cognitive and linguistic studies of the underlying functional sources of language disorders; investigations targeted at assessment and treatment of language disorders; and comparative study of language and its disorders in different populations.

Brain-Language Relations

Clearer understanding of the complex relationship between language processing and brain structure and function is essential to addressing a variety of crucial issues relating to language and its disorders in adults. Primary among these is a set of questions relating to recovery of language function. Individuals vary widely in their recovery of language capacities after apparently-similar brain lesions and little is known about the mechanisms underlying this variability. Detailed study is needed to:

- o Investigate the mechanisms by which recovery takes place and the basis for individual differences in recovery, using a variety of techniques for indexing brain activity *in vivo* (e.g., positron emission tomography, single photon emission computed tomography, event-related potentials).
- o Clarify the role of the non-dominant (typically right) cerebral hemisphere in the assumption of language functions following damage to the dominant hemisphere.
- o Relate specific symptoms to discrete brain regions using lesion analysis. A correlation between the breakdown of motor speech planning and structural brain lesions has been demonstrated, but other potentially localizable language skills (such as speech sound discrimination and oral

word comprehension) are still inadequately mapped.

- o Define by clinico-anatomic study the link between processes that are closely related to well-localized structures and those that are broadly distributed in the language zone.
- o Identify well-defined forms of selective impairments through detailed study of individual cases, including careful analysis of brain lesions and of functional language impairment.
- o Explore the role of various neurotransmitter systems in the mediation of particular language operations. The potential for pharmacological treatment follows directly from such understanding.

Analysis of Processes Underlying Language Disorders

The language impairments that result from brain damage reflect the dysfunction of one or more of the mechanisms that subserve normal language, whether spoken, written, or signed, including perceptual, motor, cognitive and specifically-linguistic mechanisms. For example, the normal ability to read involves visual perceptual mechanisms, mechanisms that control attention to the relevant material on the page, memory components that store knowledge of words and linguistic mechanisms that compute the meanings

of sentences. Damage to any one of these mechanisms will result in a reading impairment, the specific form of impairment being determined by the particular mechanism that is damaged. Thus, an understanding of the nature of the various forms of language disorders consequent to brain damage crucially depends on knowledge of normal-language and related processes and their disruption. Continued study is needed to:

- o Extend recent progress in the cognitive sciences to an understanding of the structure of normal language processing. For example, techniques involving real-time analysis of normal-language processes should be adapted to examine the nature of language pathology.
- o Distinguish the various forms of dysfunction that characterize oral/aural, written and signed languages through comparative study of impairments in those modalities.
- o Elucidate the relationship between deficits that are specific to the language system and those that arise from perceptual, motor or other cognitive disorders and clarify their relative contributions to complex language impairments. Information about such relationships could provide an important basis for the design of new treatments.

- o Continue comparative analyses of disorders in different languages, on the assumption that language-specific differences in symptoms can help reveal the linguistic and cognitive mechanisms that subserve language processing.
- o Develop artificial intelligence computer models that simulate normal language and its disorders.

Assessment, Intervention and Recovery

Approaches to the remediation of language disorders require objective and reliable information about the nature and stability of patients' symptoms. Assessment instruments that provide such information must be developed in light of new research findings that have clarified symptom and deficit relationships. Furthermore, intervention strategies to address patients' impairments should build upon emerging indications that some symptoms indicate specific functional deficits and should exploit new technologic innovations. Specific opportunities in these areas include efforts to:

- o Demonstrate the efficacy and efficiency of the existing intervention strategies for aphasia treatment. Such demonstrations will require development of new approaches to the evaluation of efficacy which use both single subject and group methods.

- o Revise and improve existing assessment procedures in order to bring them into line with current understanding of the relationship of patients' symptoms to causal underlying language deficits.
- o Develop a workable diagnostic scheme that provides a rational basis for the classification of patients and for comprehensive characterization of their deficits.
- o Develop functional assessment tools to evaluate the communicative abilities of language-impaired individuals.
- o Study the patterns of symptoms in patients with disorders in which deterioration instead of improvement is the natural course, for example, in Alzheimer's disease. Identification of predictors of the course of progression of language disorders in such patients would contribute to their management and provide a basis for approaching intervention.
- o Develop and evaluate computer-assisted instruction as an intervention strategy for working with aphasic individuals. In particular, strategies that capitalize on recent information concerning the functional basis of specific language symptoms or that provide aphasic individuals with workable compensatory mechanisms should be developed.

- o Develop and evaluate alternative and augmentative communication systems for adults with language deficits. Adults severely-impaired in understanding and producing language may be capable of effective communication by alternative means, including computer-based systems.
- o Investigate social and cultural factors that have the potential to affect treatment outcome and evaluate the relative contribution of these factors to individual differences in response to treatment.

Comparative Language Studies

Language disorders in adults can take a variety of forms, because the many causes of such disorders affect the system differently and because there are pre-existing differences among the individuals who develop language problems. This diversity of language symptoms provides an important research opportunity, as comparative study of the different forms of language impairment can provide information about the nature of underlying language mechanisms. New initiatives are needed to:

- o Obtain normative data from healthy individuals of various ages to determine whether or not there are changes throughout the life

span in the use of spoken, written or signed language.

- o Develop normative data on adults from a range of social strata and linguistic and cultural backgrounds to provide a basis for addressing the contributions of sociocultural factors, bilingualism and normal aging to language impairments.
- o Compare analysis of left- and right-brain damaged hearing subjects to determine the extent to which the right hemisphere may be involved in language use. Study brain-damaged users of sign language in order to understand hemispheric specialization for complex linguistic properties conveyed through spatial mechanisms.
- o Study the differences that exist between the language disorders of focally brain damaged patients and those that accompany more generalized brain dysfunction. Comparisons among the forms that language breakdown can take secondary to these various causes have suggested important functional dissociations, but further study is required to clarify the nature of and underlying bases for these differences.
- o Develop normative data on the language functioning of diverse populations to provide a substantive basis for approaching

their language disorders. The population of the United States is increasingly multilingual; as this population ages and its risks of stroke and consequent aphasia

increases, there will be an increasing number of bilingual individuals with language disorders.

**NATIONAL STRATEGIC RESEARCH PLAN
for
Balance and the Vestibular System
and
Language and Language Impairments
1991**

ERRATA

APPENDIX A - Public Law 100-553

Pages are reversed.

Change page 105 to 104
104 to 105
107 to 106
106 to 107

APPENDIX A

Public Law 100-553

Public Law 100-553
100th Congress

An Act

To amend the Public Health Service Act to establish within the National Institutes of Health a National Institute on Deafness and Other Communication Disorders.

Oct. 28, 1988
(S. 1727)

Be it enacted by the Senate and House of Representatives of the United States of America in Congress assembled,

SECTION 1. SHORT TITLE.

This Act shall be cited as the "National Deafness and Other Communication Disorders Act of 1988".

National
Deafness and
Other
Communication
Disorders Act of
1988.
42 USC 201 note.

SEC. 2. ESTABLISHMENT AND TRANSFER OF FUNCTIONS.

Title IV of the Public Health Service Act (42 U.S.C. 281 et seq.) is amended—

(1) in section 401(b)(1)—

(A) by striking "and Communicative" in subparagraph (J); and

(B) by adding at the end the following new subparagraph:
"(M) The National Institute on Deafness and Other Communication Disorders.";

(2) in the heading for subpart 10 of part C, by striking "and Communicative";

(3) in section 457—

(A) by striking "and Communicative"; and

(B) by striking "disorder, stroke," and all that follows and inserting "and disorder and stroke."; and

(4) in Part C, by adding at the end the following new subpart:

"Subpart 13—National Institute on Deafness and Other Communication Disorders

Education.
Research and
development.
Public
information.

"PURPOSE OF THE INSTITUTE

"Sec. 464. The general purpose of the National Institute on Deafness and Other Communication Disorders (hereafter referred to in this subpart as the 'Institute') is the conduct and support of research and training, the dissemination of health information, and other programs with respect to disorders of hearing and other communication processes, including diseases affecting hearing, balance, voice, speech, language, taste, and smell.

42 USC 286m.

"NATIONAL DEAFNESS AND OTHER COMMUNICATION DISORDERS PROGRAM

"Sec. 464A. (a) The Director of the Institute, with the advice of the Institute's advisory council, shall establish a National Deafness and Other Communication Disorders Program (hereafter in this section referred to as the 'Program'). The Director or the Institute shall, with respect to the Program, prepare and transmit to the Director of NIH a plan to initiate, expand, intensify and coordinate activities of the Institute respecting disorders of hearing (including tinnitus) and

42 USC 286m-1.

"MULTIPURPOSE DEAFNESS AND OTHER COMMUNICATION DISORDERS
CENTER

"Sec. 464C. (a) The Director of the Institute shall, after consultation with the advisory council for the Institute, provide for the development, modernization, and operation (including care required for research) of new and existing centers for studies of disorders of hearing and other communication processes. For purposes of this section, the term 'modernization' means the alteration, remodeling, improvement, expansion, and repair of existing buildings and the provision of equipment for such buildings to the extent necessary to make them suitable for use as centers described in the preceding sentence.

42 USC 285m-3.

"(b) Each center assisted under this section shall—

"(1) use the facilities of a single institution or a consortium of cooperating institutions; and

"(2) meet such qualifications as may be prescribed by the Secretary.

"(c) Each center assisted under this section shall, at least, conduct—

"(1) basic and clinical research into the cause diagnosis, early detection, prevention, control and treatment of disorders of hearing and other communication processes and complications resulting from such disorders, including research into rehabilitative aids, implantable biomaterials, auditory speech processors, speech production devices, and other otolaryngologic procedures;

"(2) training programs for physicians, scientists, and other health and allied health professionals;

Health care
professionals.

"(3) information and continuing education programs for physicians and other health and allied health professionals who will provide care for patients with disorders of hearing or other communication processes; and

"(4) programs for the dissemination to the general public of information—

"(A) on the importance of early detection of disorders of hearing and other communication processes, of seeking prompt treatment, rehabilitation, and of following an appropriate regimen; and

"(B) on the importance of avoiding exposure to noise and other environmental toxic agents that may affect disorders of hearing or other communication processes.

"(d) A center may use funds provided under subsection (a) to provide stipends for health professionals enrolled in training programs described in subsection (c)(2).

"(e) Each center assisted under this section may conduct programs—

"(1) to establish the effectiveness of new and improved methods of detection, referral, and diagnosis of individuals at risk of developing disorders of hearing or other communication processes; and

"(2) to disseminate the results of research, screening, and other activities, and develop means of standardizing patient data and recordkeeping.

"(f) The Director of the Institute shall, to the extent practicable, provide for an equitable geographical distribution of centers assisted under this section. The Director shall give appropriate consideration

Aged persons.
Children and
youth.

other communication processes, including diseases affecting hearing, balance, voice, speech, language, taste, and smell. The plan shall include such comments and recommendations as the Director of the Institute determines appropriate. The Director of the Institute shall periodically review and revise the plan and shall transmit any revisions of the plan to the Director of NIH.

"(b) Activities under the Program shall include—

"(1) investigation into the etiology, pathology, detection, treatment, and prevention of all forms of disorders of hearing and other communication processes, primarily through the support of basic research in such areas as anatomy, audiology, biochemistry, bioengineering, epidemiology, genetics, immunology, microbiology, molecular biology, the neurosciences, otolaryngology, psychology, pharmacology, physiology, speech and language pathology, and any other scientific disciplines that can contribute important knowledge to the understanding and elimination of disorders of hearing and other communication processes;

"(2) research into the evaluation of techniques (including surgical, medical, and behavioral approaches) and devices (including hearing aids, implanted auditory and nonsensory prosthetic devices and other communication aids) used in diagnosis, treatment, rehabilitation, and prevention of disorders of hearing and other communication processes;

"(3) research into prevention, and early detection and diagnosis, of hearing loss and speech and language disturbances (including stuttering) and research into preventing the effects of such disorders on learning and learning disabilities with extension of programs for appropriate referral and rehabilitation;

"(4) research into the detection, treatment, and prevention of disorders of hearing and other communication processes in the growing elderly population with extension of rehabilitative programs to ensure continued effective communication skills in such population;

"(5) research to expand knowledge of the effects of environmental agents that influence hearing or other communication processes; and

"(6) developing and facilitating intramural programs on clinical and fundamental aspects of disorders of hearing and all other communication processes.

"DATA SYSTEM AND INFORMATION CLEARINGHOUSE

42 USC 285m-2.

"Sec. 464B. (a) The Director of the Institute shall establish a National Deafness and Other Communication Disorders Data System for the collection, storage, analysis, retrieval, and dissemination of data derived from patient populations with disorders of hearing or other communication processes, including where possible, data involving general populations for the purpose of identifying individuals at risk of developing such disorders.

"(b) The Director of the Institute shall establish a National Deafness and Other Communication Disorders Information Clearinghouse to facilitate and enhance, through the effective dissemination of information, knowledge and understanding of disorders of hearing and other communication processes by health professionals, patients, industry, and the public.

the annual rate in effect for grade GS-18 of the General Schedule for each day (including traveltime) they are engaged in the performance of their duties as members of the Board.

"(d) The term of office of an appointed member of the Advisory Board is four years, except that no term of office may extend beyond the expiration of the Advisory Board. Any member appointed to fill a vacancy for an unexpired term shall be appointed for the remainder of such term. A member may serve after the expiration of the member's term until a successor has taken office. If a vacancy occurs in the Advisory Board, the Secretary shall make an appointment to fill the vacancy not later than 90 days from the date the vacancy occurred.

"(e) The members of the Advisory Board shall select a chairman from among the appointed members.

"(f) The Secretary shall, after consultation with and consideration of the recommendations of the Advisory Board, provide the Advisory Board with an executive director and one other professional staff member. In addition, the Secretary shall, after consultation with and consideration of the recommendations of the Advisory Board, provide the Advisory Board with such additional professional staff members, such clerical staff members, such services of consultants, such information, and (through contracts or other arrangements) such administrative support services and facilities, as the Secretary determines are necessary for the Advisory Board to carry out its functions.

"(g) The Advisory Board shall meet at the call of the chairman or upon request of the Director of the Institute, but not less often than four times a year.

"(h) The Advisory Board shall—

"(1) review and evaluate the implementation of the plan prepared under section 464A(a) and periodically update the plan to ensure its continuing relevance;

"(2) for the purpose of assuring the most effective use and organization of resources respecting deafness and other communication disorders, advise and make recommendations to the Congress, the Secretary, the Director of NIH, the Director of the Institute, and the heads of other appropriate Federal agencies for the implementation and revision of such plan; and

"(3) maintain liaison with other advisory bodies related to Federal agencies involved in the implementation of such plan and with key non-Federal entities involved in activities affecting the control of such disorders.

"(i) In carrying out its functions, the Advisory Board may establish subcommittees, convene workshops and conferences, and collect data. Such subcommittees may be composed of Advisory Board members and nonmember consultants with expertise in the particular area addressed by such subcommittees. The subcommittees may hold such meetings as are necessary to enable them to carry out their activities.

"(j) The Advisory Board shall prepare an annual report for the Secretary which—

"(1) describes the Advisory Board's activities in the fiscal year for which the report is made;

"(2) describes and evaluates the progress made in such fiscal year in research, treatment, education, and training with respect to the deafness and other communication disorders;

Reports.

to the need for centers especially suited to meeting the needs of the elderly, and of children (particularly with respect to their education and training), affected by disorders of hearing or other communication processes.

"(g) Support of a center under this section may be for a period not to exceed seven years. Such period may be extended by the Director of the Institute for one or more additional periods of not more than five years if the operations of such center have been reviewed by an appropriate technical and scientific peer review group established by the Director, with the advice of the Institute's advisory council, if such group has recommended to the Director that such period should be extended.

**"NATIONAL INSTITUTE ON DEAFNESS AND OTHER COMMUNICATION
DISORDERS ADVISORY BOARD**

42 USC 285m-4.

"Sec. 464D. (a) The Secretary shall establish in the Institute the National Deafness and Other Communication Disorders Advisory Board (hereafter in this section referred to as the 'Advisory Board').

"(b) The Advisory Board shall be composed of eighteen appointed members and nonvoting ex officio members as follows:

"(1) The Secretary shall appoint—

"(A) twelve members from individuals who are scientists, physicians, and other health and rehabilitation professionals, who are not officers or employees of the United States, and who represent the specialties and disciplines relevant to deafness and other communication disorders, including not less than two persons with a communication disorder; and

"(B) six members from the general public who are knowledgeable with respect to such disorders, including not less than one person with a communication disorder and not less than one person who is a parent of an individual with such a disorder.

Of the appointed members, not less than five shall by virtue of training or experience be knowledgeable in diagnoses and rehabilitation of communication disorders, education of the hearing, speech, or language impaired, public health, public information, community program development, occupational hazards to communications senses, or the aging process.

"(2) The following shall be ex officio members of each Advisory Board:

"(A) The Assistant Secretary for Health, the Director of NIH, the Director of the National Institute on Deafness and Other Communication Disorders, the Director of the Centers for Disease Control, the Chief Medical Director of the Veterans' Administration, and the Assistant Secretary of Defense for Health Affairs (or the designees of such officers).

"(B) Such other officers and employees of the United States as the Secretary determines necessary for the Advisory Board to carry out its functions.

"(c) Members of an Advisory Board who are officers or employees of the Federal Government shall serve as members of the Advisory Board without compensation in addition to that received in their regular public employment. Other members of the Board shall receive compensation at rates not to exceed the daily equivalent of

"(3) summarizes and analyzes expenditures made by the Federal Government for activities respecting such disorders in such fiscal year; and

"(4) contains the Advisory Board's recommendations (if any) for changes in the plan prepared under section 464A(a).

"(k) The National Deafness and Other Communication Disorders Advisory Board shall be established not later than 90 days after the date of the enactment of the National Institute on Deafness and Other Communication Disorders Act.

"INTERAGENCY COORDINATING COMMITTEE

42 USC 285m-5.

"Sec. 464E. (a) The Secretary may establish a committee to be known as the Deafness and Other Communication Disorders Interagency Coordinating Committee (hereafter in this section referred to as the 'Coordinating Committee').

"(b) The Coordinating Committee shall, with respect to deafness and other communication disorders—

"(1) provide for the coordination of the activities of the national research institutes; and

"(2) coordinate the aspects of all Federal health programs and activities relating to deafness and other communication disorders in order to assure the adequacy and technical soundness of such programs and activities and in order to provide for the full communication and exchange of information necessary to maintain adequate coordination of such programs and activities.

"(c) The Coordinating Committee shall be composed of the directors of each of the national research institutes and divisions involved in research with respect to deafness and other communication disorders and representatives of all other Federal departments and agencies whose programs involve health functions or responsibilities relevant to deafness and other communication disorders.

"(d) The Committee shall be chaired by the Director of NIH (or the designee of the Director). The Committee shall meet at the call of the chair, but not less often than four times a year.

Reports.

"(e) Not later than 120 days after the end of each fiscal year, the Committee shall prepare and transmit to the Secretary, the Director of NIH, the Director of the Institute, and the advisory council for the Institute a report detailing the activities of the Committee in such fiscal year in carrying out subsection (b).

"LIMITATION ON ADMINISTRATIVE EXPENSES

42 USC 285m-6.

"Sec. 464F. With respect to amounts appropriated for a fiscal year for the National Institutes of Health, the limitation established in section 408(b)(1) on the expenditure of such amounts for administrative expenses shall apply to administrative expenses of the National Institute on Deafness and Other Communication Disorders."

42 USC 285m
note.
Gifts and
property.
Contracts.
Records.

SEC. 3. TRANSITIONAL AND SAVINGS PROVISIONS.

(a) **TRANSFER OF PERSONNEL, ASSETS, AND LIABILITIES.**—Personnel employed by the National Institutes of Health in connection with the functions vested under section 2 in the Director of the National Institute on Deafness and Other Communication Disorders, and assets, property, contracts, liabilities, records, unexpended balances of appropriations, authorizations, allocations, and other funds of the National Institutes of Health, arising from or employed, held, used,

available to, or to be made available, in connection with such functions shall be transferred to the Director for appropriate allocation. Unexpended funds transferred under this subsection shall be used only for the purposes for which the funds were originally authorized and appropriated.

(b) SAVINGS PROVISIONS.—With respect to functions vested under section 1 in the Director of the National Institute on Deafness and Other Communication Disorders, all orders, rules, regulations, grants, contracts, certificates, licenses, privileges, and other determinations, actions, or official documents, that have been issued, made, granted, or allowed to become effective, and that are effective on the date of the enactment of this Act, shall continue in effect according to their terms unless changed pursuant to law.

Grants.
Contracts.

Approved October 28, 1988.

LEGISLATIVE HISTORY—S. 1727 (H.R. 3361):

HOUSE REPORTS: No. 100-761 accompanying H.R. 3361 (Comm. on Energy and Commerce).

CONGRESSIONAL RECORD, Vol. 134 (1988):

Oct. 7, considered and passed Senate.

Oct. 13, considered and passed House.

APPENDIX B

**Balance and the
Vestibular System**

and

**Language and
Language Impairment**

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APPENDIX B

Resource Persons

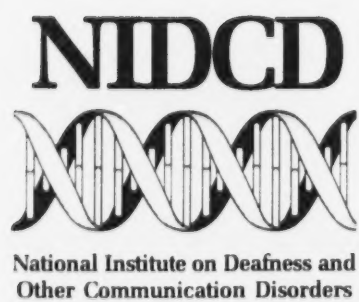
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